THE BIRTH OF A CHILD WITH A CONGENITAL ANOMALY: SOME PSYCHOSOCIAL IMPLICATIONS FOR THE FAMILY

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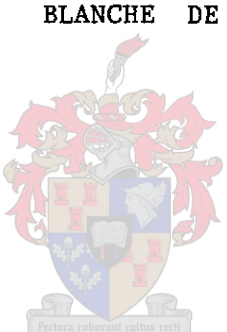
by

BLANCHE DE WET



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CHAPTER 1

INTRODUCTION

A. INTRODUCTION

During the 5 year period 1979 through 1983 between 140 and 200 (average 178) patients with birth defects were admitted each year to Red Cross War Memorial Children's Hospital, Cape Town* for corrective surgery. The congenital anomalies seen differ widely both in severity and in the extent to which surgical procedures can effect restoration to normal physical functioning. Routine documentation of procedures and outcome facilitates regular evaluation and ensures further action in those cases where the fullest physical potential has not yet been reached. At the same time research projects geared primarily towards the improvement of treatment techniques are constantly in progress.

As yet very little is known however about the way in which the families of these patients respond to the inherent problems and difficulties and their attempts to overcome them. In this respect no systematic documentation exists which can lead to valid evaluation, followed by appropriate action. Although an increasingly good quality of physical life is therefore becoming possible for patients with birth defects, knowledge of the outcome for their families accompanied by constructive methods of ongoing support, have not accumulated and developed concurrently.

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* For the purposes of this study alternatively referred to as the Children's Hospital.

The need for empirical research in this respect becomes even more apparent when it is realized that patients with identical surgical results, often show divergent outcomes in residual symptoms and adjustment. Undoubtably the patient's difficulties have some effect on the family but at the same time the different coping strategies of families seem to have a bearing on the patient's physical well-being. It is clear that the patient and his anomaly could not be seen in isolation to his family and that a very real need exists to work towards bringing knowledge of these factors on a par with knowledge of surgical factors.

The need to study the family of the sick or handicapped child has not gone unrecognized in the literature. Especially since the 1960's, many articles from many different disciplines have been published looking at the family and the handicapped in general (Baum,1962; Burden,1978,1980; Tavormina,1981; Younghusband et al, 1970; Fox,1975; Lonsdale,1978; MacKeith,1973; Poznanski,1973) longterm childhood illness and the family (Mattson,1972; Sigal et al, 1973; Travis,1976) and the family and patient birth defects in general (Drotar et al,1975; Gur,1974; Griffith,1974; Tew et al, 1974). Some researchers (Burton, 1976; Begleiter,1976; Oppenheimer and Rucker, 1980; and Mittenzwey,1981) have studied the family and a specific inherited disease like cystic fibrosis or a birth defect like Spina Bifida (Dorner,1975; Freeston,1971; Joosten,1979; Seidler,1981) to name a few. Still some other authors have dealt with the moral and ethical issues of selection for treatment (Zachary,1968; De Lange,1974; Lorber,1974;Katzen,1981; Ackerman, 1980) and whether active management could be justified in certain cases (Cywes,1980; Eckstein,1979; Duff,1981). Although varied

approaches have been used by these authors, the common theme which emerges is firstly a recognition of the fact that successful coping of the family is of crucial importance to the affected child's well-being. Secondly, there is consensus that the physical well-being of the patient is of dubious value if the emotional survival of the family were at stake.

The foregoing stresses the essential need to know more about the families of patients if treatment results are to be maximised. Questions like the following need to be answered:
For which proportion of families is the emotional survival being threatened by the patient's anomaly and the attendant demands?
Does the proportion differ favourably for those cases where it was possible to totally correct an anomaly through surgery?
Do the problems posed to the family by different anomalies differ fundamentally?
What are the coping strategies employed by families and which factors hinder or enhance these efforts?

Some of these questions have been dealt with in the literature. Certain anomalies, notably cystic fibrosis, have often been documented in this respect, especially in the literature of the United Kingdom and the United States of America. In contrast other anomalies like Hirschsprung's disease, oesophageal atresia and anorectal malformations which are seen far more frequently in the Department of Paediatric Surgery of the Children's Hospital, have hardly,if ever,been researched in this connection.

The present study is a tentative first step to fill this void and to find answers to the foregoing questions.

B. THE AIM OF THE STUDY

In general terms the aim of this study is to gain insight into the psychosocial difficulties of families where children with life-threatening birth defects requiring surgical intervention viz. oesophageal atresia, Hirschsprung's disease or anorectal malformations are born as compared with an hereditary condition like cystic fibrosis.

1. It is firstly hypothesized that a smaller proportion of the families of patients with life-threatening surgical conditions will have been aided by the services of social workers than the families of cystic fibrosis patients.
2. It is secondly hypothesized that a common core of difficulties would be present in all four of the diagnostic categories studied, while other difficulties would be peculiar to a specific type of anomaly only, with its own phases of high risk for the families involved.
3. It is thirdly expected that the duration of psychosocial difficulties related to the anomaly will be most limited for the families of those patients where an anomaly was corrected, leaving the minimum of residual symptoms.

The specific aims of the study are therefore:

1. To make a systematic investigation into the nature of demands upon the family posed by the birth of a child with Hirschsprung's disease, cystic fibrosis, anorectal malformations, and oesophageal atresia;

2. To determine the specific difficulties inherent to each anomaly and to isolate the high risk phases for each.
3. To make recommendations for enhancing the coping strategies of patients and their families, with the view to social work intervention specifically geared to the identified needs.

C. METHODS

With the foregoing aims in mind, the researcher is faced with the question of selecting an appropriate research design as well as finding a theoretical model to serve as a basis for the study. The basis of selection utilized in this study is briefly highlighted, followed by a description of the practical implementation thereof.

1. The choice of a research design

The review literature has been unrelenting in its criticism of almost any type of research design which has attempted to study the effects of a variety of variables on the family. Frequently mentioned shortcomings have included biased sampling (Frydman,1970), subjective evaluations (Lewis and Khaw, 1982), clinical impressions (Gayton et al, 1977) and a lack of comparison groups (Drotar et al, 1981). In certain instances this critique is well founded, as many researchers have failed to deal scientifically with the material studied. In many cases, however, reality factors have precluded the use of a more rigorous methodology. All the same, caution should be exercised before summarily rejecting certain methods in favour of other research designs in an effort to obtain more valid results.

Chess and Thomas (1982) warned that the much acclaimed experimental design holds the danger of "... transposing findings from artificial experimental settings to real life situations"(p.219). Mc Call (1977) reasoned that the experimental method "... dictates rather than serves ..." the problems addressed in developmental studies and asks the question: "What value is our knowledge if it is not relevant to real children growing up in real families and in real neighbourhoods?" (p.334). Kitson et al (1982) in their discussion of sampling issues in family research, argued that control groups could well enhance areas in this field, but point out the following restrictions: "...it is typically possible to compare the two (or more) groups on only a limited number of characteristics. Other characteristics that were not measured or were not included in the definition of the study population may produce systematic biases that affect the findings, with the result that differences between the groups may be due to differences in sample characteristics, not differences due to the intervention or change in the characteristics of interest" (p.972).

Some researchers have in fact found that tests chosen to measure certain effects on the family, were not valid for the particular set of circumstances which they were meant to be studying. Thus Miller (1973) found that although parents of children with congenital heart defects,were expressing anxiety both verbally and non-verbally,this expressed anxiety did not correlate with the actual test findings, which reflected their anxiety as being within the normal to low range. On this point she concluded that the indications were that " ... the test used is not valid to measure a transitional, situationally-induced state of anxiety"

(Miller, 1973,p.39). The question arises as to what extent researchers have succeeded in selecting and finding instruments which are appropriate and sensitive enough to measure areas which might reflect effects on the family of the child with an anomaly.

Judson and Burden (1980) adequately described the many difficulties in finding relevent criteria by which to evaluate change in parental attitude, in spite of the fact that a wide range of instruments were available for this purpose. "However it is well known that one only gets answers to the questions asked and inevitably, most of the available instruments will be blunt in terms of sensitivity to the changes anticipated from a particular project" (p.49). In their review of research on families in crisis pertinent to mental retardation in children, Jacobson and Humphrey (1979) point out that where procedures are designed to measure an assumed negative effect, it "... may unknowingly direct attention away from any positive family results" (p.601).

The comments of the aforementioned authors have clearly demonstrated that a pre-requisite for this type of methodology would be the existence of a sound and extended knowledge of the material under study. A similar problem is faced when the researcher attempts to avoid the criticisms so often levelled at the retrospective study (i.e. selective memory and distortion) as prospective studies equally require an extended knowledge of the subject material. "If a significant factor is overlooked in a prospective study it cannot be retrieved" (Stott, 1973 p.770). The solution to this problem offered by Stott probably holds true for the larger part of research designs

discussed so far: "The most sophisticated strategy, over the long term, is to carry out retrospective studies of selected conditions in order to make the initial discoveries, ... which form the hypotheses for testing by prospective investigation" (p.770). The questionnaire, which is one of the most frequently used techniques for making such "initial discoveries", is not above censure. In her search for suitable research tools to investigate family life with a handicapped child, Voysey (1975) notes that "... questionnaires in effect filter the social processes under study through a pre-defined 'grid' of categories assumed to represent the range of possible alternative responses appropriate to the area of research..." of which she felt fixed choice questions represented the extreme. "It is meaningless to produce measurements or quantifications of phenomena whose dynamics are not yet understood"(p.66).

In their methodological overview of research on health care and the family, Litman and Venters (1979) point out that longitudinal studies, for all their advantages, are not without inherent difficulties. "Foremost amongst these is the rather high rate of attrition and non-response" (p.380). Apart from prohibitive financial and organisational costs, "... very long projects may be subject to premature termination or incompleteness due to the unanticipated death of either the subjects and/or the investigator" (p.380). To circumvent these dilemmas, a limited number of authors have innovated techniques of intergenerational analyses of health problems and the family.

In the face of such formidable methodological issues it is hardly surprising that researchers have recently examined again the means for more rigorous application of the case study method (Runyan,1982) and how to improve its internal validity (Kazdin, 1981). The familiar argument that this method can be misused to apply the researcher's theoretical preferences in an arbitrary fashion, elicited the comment that this undoubtedly happens at times, "... but any method can be poorly used. The fact that one can lie with statistics or can misuse the case study method is no reason to abandon the method, but rather implies that it needs to be used with proper caution and controls" (Runyan 1982 p.444).

In view of the foregoing and the never ending search for suitable research designs, the conclusions of meticulous methodologists like Hirschi and Selvin and Kitson and her associates are well remembered: "We should not be so quick to judge severely and to negate research results obtained through a variety of sampling methods; rather, we should discuss them in terms of their assets and liabilities. We need to accept studies on their merits, however limited they are, and build upon them through replication" (Kitson et al,1982). "In the end, therefore, the methodologist should be sympathetic enough to recognise that everyone makes mistakes, vigilant enough to guard against taking the occasional error as an indicator of overall quality, and objective enough to understand that the most visible target is provided by the best research. In return he can hope that other methodological critics will appraise his work in the same spirit" (Hirschi and Selvin, 1973). It is trusted that the eclectic research design chosen for the present study (see pp.26-37), attempting to describe the

realities faced by our families, will be judged in the light of the many difficulties facing the researcher studying the family.

2. A conceptual framework for studying the family of the child with a congenital anomaly

The lack of utilizing consistent, integrated theory or a working model as a basis for research on health and the family, has been lamented by various authors for many years (Riskin, 1963; Minde et al,1972; Litman and Venters, 1979). The latter authors have adequately described the theoretical and conceptual aspects of family health care research. Probably the most frequent attempts at model building and testing by means of hypotheses have come from the field of psychiatry and psychology. Thus Minde et al (1972) sought to assess the validity of three models expounded by others and Chess et al (1980) illustrated the concepts of goodness of fit and consonance - dissonance in the developmental process of the handicapped child in the family, to name but a few.

A pertinent area which has certainly seen replication, testing and modification is found in life events research. Where previous findings have indicated some association between the crowding of life events and the onset of illness, Holmes and Rahe (1967) set out to determine the magnitude of life change brought about by these different life events. Paykel et al (1969) in a controlled study endorsed the importance of such life events in the onset of depression. Furthermore undesirable events and those comprising losses or exits from the social field were isolated by the latter

authors as especially relevant. In 1975 Vinokur and Selzer confirmed that, "... only the undesirable events were substantially correlated with the stress related variables and that these events required greater adjustments than the desirable ones" (p.335). This was soon followed by the construction of a scale to measure the stress of life events, which included the concepts of both Holmes and Paykel (Tennant and Andrews, 1976). Life graph findings, a variation of the foregoing, indicated that events "... seem to be measured against an implicit schedule according to which they are seen as traumatic or acceptable" (p.669). Although life events research has been criticized for ascribing an almost linear relationship between life events and illness (Rabkin and Struening,1976) it has nevertheless yielded valuable results with a possible contribution to understanding the family of the child with a congenital anomaly. Insofar as parents interpret the anomaly as undesirable, with the added unexpected nature of the defect and the loss of the normal child they had hoped for, this can undoubtedly be defined as a major life event. Three of the vital concepts requiring major readjustment i.e. undesirability, unexpectedness and loss, are present when a child with a congenital anomaly is born. In terms of life events research, considerable life stress can be expected in such families, and viewing it as a crisis would not be alien to the literature studied.

The conceptual framework "... most used by family sociologists in the study of family crisis" (p.139) outlined by Reuben Hill (1958), was found to be the most comprehensive as a baseline for the present study. As social workers should be familiar with these

concepts, they will only be briefly highlighted here and applicable aspects referred to in more detail in the empirical study.

In this ABCX model, A, the event or stressor, interacts with B, the family's crisis-meeting resources, which in turn reacts with C, the family's interpretation or definition of the event, which produces X, the crisis. The stressor is defined as "... a situation for which the family has had little or no prior preparation and must therefore be viewed as problematic"(p.140). The impact of a specific stressor can never be the same for any given family, as the hardships which accompany it will never be exactly similar. (Thus the material hardships of families geographically distant from a hospital may be more severe than for those in close proximity, although the stressor or anomaly of the child may be similar.) Hill states that these hardships lie outside the family as part of the stressor, while family resources and the meaning attached to the stress event, lie within the family itself, closely related to its structure and values. A stressor can only become a crisis by the meaning attached to it by the family. The "meaning" dimension therefore explains the different reactions of families to the same event or stressor.

Two of the classifications of stressor events offered by Hill are pertinent to the present study.

(a) Source of trouble

The source of trouble can either be extra or intra-familial. Where

stressors are outside, or beyond control of the family (viz economic depression) family solidarity may even increase. Stressors originating from interpersonal relationships within the family itself (viz infidelity or alcoholism) are more likely to lead to family disorganisation and breakdown. In the context of the child with a congenital anomaly it would clearly be important to establish whether parents blame one another (stressor becomes intra-familial), or whether it is regarded as a chance event which could have happened to any family (stressor becomes extra-familial).

(b) Effect on family configuration

The second type of classification involves loss of a family member (dismemberment) or the addition of any unprepared-for member (accession) and loss of family morale or unity (demoralization). Hill includes stressors which require major role changes, although the plurality of the family may remain unchanged. The applicability to the study of the family where a child with an anomaly is born, is selfapparent.

The family's crisis meeting resources (the B factor) go beyond social class and material means to include vital aspects such as family integration and family adaptability. These are called upon mainly because crises tend to inert or delay emotionally-satisfying behaviour while at the same time demanding major shifts in roles of the individual family members. In the family of a child with a congenital anomaly, such role shifts may become semi-permanent, due

to the prolonged nature of the stressor.

In conclusion it needs to be stressed that the "meaning" dimension can transcend available resources. "Not infrequently families with objective resources adequate to meet the hardships of sickness or job loss crack under stress because they define such hardship situations as "insurmountable"(p.145). The present empirical study attempted to gain a better understanding of the C-factor, i.e. the meaning dimension of Hill's ABCX model of studying families under stress. For this reason every attempt was made to bring the parent's interpretation of the birth of a child with congenital anomaly across. In this respect citations from case histories will be extensively used. The motivation was that understanding the C-factor would be the most crucial variable in successful social work intervention with these families. More distinctly spelt out: Controlling the A-factor or stressor (in this context changing or removing the anomaly) does not fall within the scope of social work intervention. Furthermore, as research results indicated, the B-factor or crisis-meeting resources of families varied extensively. Yet the materially well-equipped families, viz. social classes I and II, (see Chapter 4) did not necessarily experience less difficulty following the birth of a child with a congenital anomaly. Therefore looking closely at the meaning which parents attach to this event, assumes principal importance in understanding their difficulties.

3. Literature search and pilot study

A literature search was done with the aid of inter alia the

Institute of Medical Literature of the South African Medical Research Council to serve as a basis for the empirical study. Because of the paucity of relevant material relating to oesophageal atresia, anorectal malformations and Hirschsprung's disease, a pilot study with 35 respondents in all the diagnostic categories was done to further guide the content of the interview schedule of the empirical study. The main findings from the literature are presented in Chapter 2 and 3.

4. DEMARCATION OF THE STUDY

The population of study was limited to all white admissions to the Children's Hospital between 1961 and 1980, who were resident in the Republic of South Africa or South West Africa at the time of study, where a diagnosis of Hirschsprung's disease, cystic fibrosis, anorectal malformation, or oesophageal atresia had been made. This included patients who had been diagnosed at other hospitals, but continued treatment at the Children's Hospital.

The rationale for excluding other race groups was firstly to narrow down cultural differences in handling of, and attitude towards congenital anomalies. Secondly it was found that black patients very often lived within an extended family where support systems were markedly different from those of the nuclear family and would therefore warrant a separate study.

The decision to study the four abovenamed anomalies was firstly based on the fact that a substantial number of families in the overall population are involved (See Chapters 2 and 3). Secondly

the lack of psychosocial follow-up results in the case of oesophageal atresia, Hirschsprung's disease and anorectal malformations indicated the need for knowledge in this area. Furthermore cystic fibrosis has been well documented which served as a basis of orientation for the present study. Cystic fibrosis, was at the same time not correctable, but hereditary and not always noticeable at birth. The other three anomalies were all operable with varying degrees of success, with oesophageal atresia probably having the most favourable prognosis after surgery. The assumption was that the inherent differences in diagnosis, treatment and residual symptoms (described in Chapters 2 and 3) would not only form a useful basis for comparison for the aims previously set out, but also facilitate selective extrapolation to other comparable conditions.

5. THE STUDY SAMPLE

(a) Selection of the sample

According to the criteria described earlier, a hospital computer printout of admissions was checked against doctors' summaries of patients with the above mentioned conditions for possible omissions. This sample frame yielded a study population of 247. Using the latter as a primary sampling unit, a random sample of 83 was drawn and patient tracing commenced. Although a heavy investment in time, money and effort was made mainly because of distances involved, 13 of the selected patients could not be traced. Due to the time consuming nature of tracing, field work was done, as soon as reliable addresses became available. In the end replacements of untraceable patients were made by substitutes

from the sample population matched for diagnosis, age and sex. Affected siblings of patients were included in the sample in cases where they also formed part of the study population to allow recording of similarities and differences in response within the same family. A final sample of 90 patients in 83 families was studied.

(b) Representativeness of the sample

Representativeness of the sample was statistically tested on two factors namely the proportion of patients in each diagnostic category and the male to female ratio within each diagnosis. Table 1 shows the diagnosis of patients in the study sample compared to the study population. Both groups were proportionately the same.

TABLE 1

DIAGNOSIS OF STUDY SAMPLE COMPARED TO STUDY POPULATION

Anomalies	Study Sample N	Study Population N	Total N
Hirschsprung's Disease	30	70	100
Cystic Fibrosis	17	49	66
Anorectal Malformations	20	54	74
Oesophageal Atresia	23	74	97
TOTAL	90	247	337

X^2 = 1.0330 P> .05: not significant
D.F. = 3

In the composite table 2 the gender ratio of patients by diagnosis is compared between the sample and the study population.

TABLE 2

GENDER RATIO OF PATIENTS BY DIAGNOSIS: SAMPLE AND STUDY POPULATION COMPARED

Hirschsprung's disease	Males	Females	Total
Study Sample	23	7	30
Study Population	55	15	70
TOTAL	78	22	100

X^2 = .0444

Cystic fibrosis	Males	Females	Total
Study Sample	8	9	17
Study Population	23	26	49
TOTAL	31	35	66

X^2 = .00007

Anorectal malformations	Males	Females	Total
Study Sample	12	8	20
Study Population	32	22	54
TOTAL	44	30	74

X^2 = .003322

Oesophageal atresia	Males	Females	Total
Study Sample	12	11	23
Study Population	38	36	74
TOTAL	50	47	97

X^2 = .0047533

The foregoing shows that the proportion of males and females within a specific diagnosis in the sample, did not differ statistically significantly from that of the study population. This was true of all diagnoses.

(c) Sample bias

Other researchers have noted that fruitless efforts in tracing large proportions of families may well reflect mobility due to crisis (De Frain and Ernst, 1978) or might result in underrepresentation of social classes 4 and 5 (Dorner, 1975). Many authors have criticized research design, including poor sampling (Stott,1973; Halstead 1976;Cummings Bayley and Rie 1966) in retrospective studies of this nature, yet no constructive alternatives to bridge these problems have been forthcoming.

In the present study it was initially assumed that bias could be towards an over-representation of those patients with residual symptoms because of a greater willingness in such cases to respond to tracing efforts. Where no problems were experienced, parents may well have argued that they could not really contribute to research, as was reported in certain instances. However cases on the other extreme where family disintegration or total abandonement of the patient resulted, proved equally difficult to contact, even with the aid of community based welfare agencies. From the discussion in Chapter 4 it will become clear that social classes 4 and 5 were also within the normal range in the present sample. It was therefore finally concluded that bias would be towards an under-representation of the two extremes: families experiencing

little or no problems and those with severe difficulties. As a 36% sample was drawn, this possible error need not be regarded as serious. A definite under representation of cases where patients died soon after birth, resulted due to parents' discontinued contact with the hospital and the ensuing tracing difficulties.

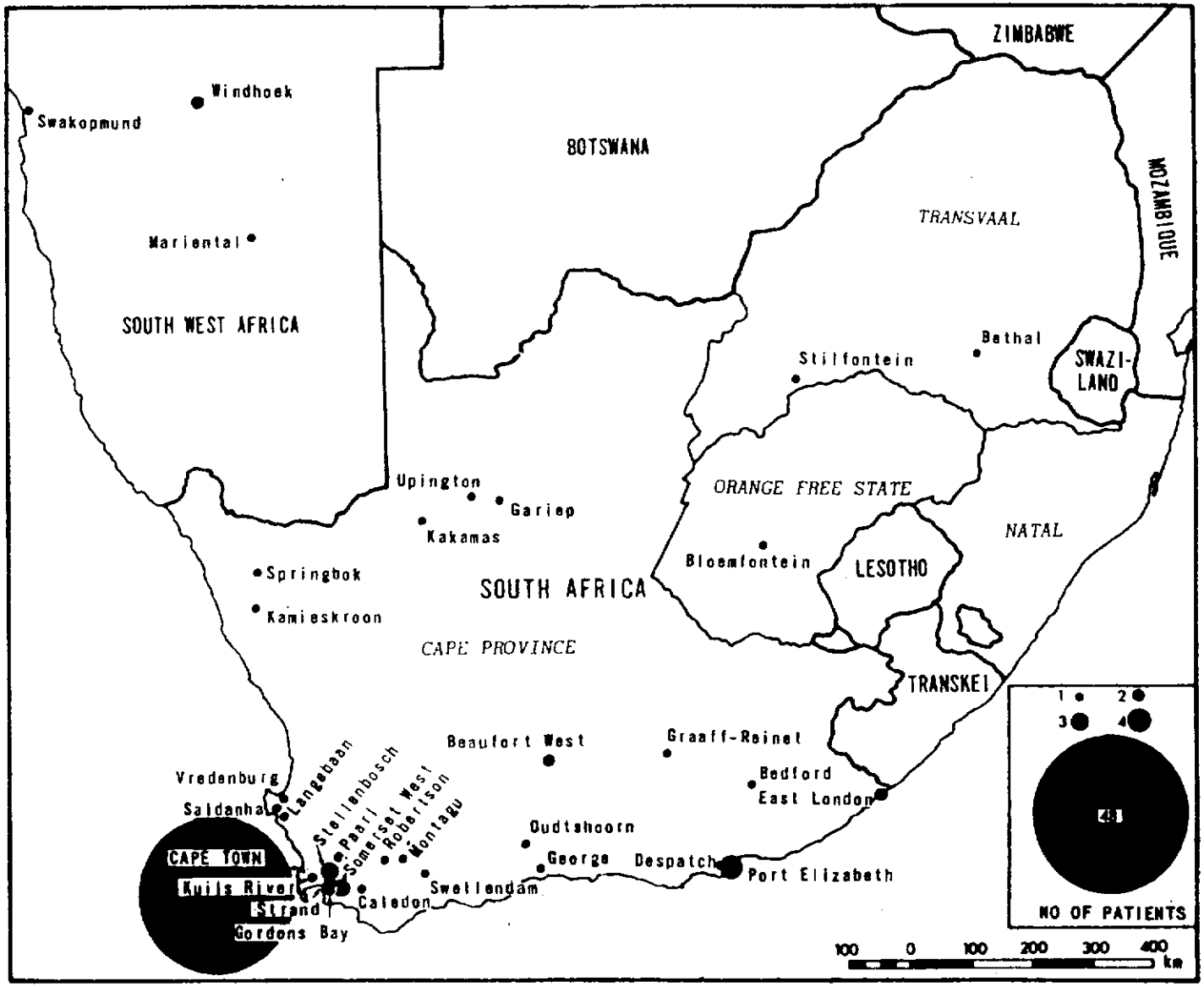
(d) Geographical distribution of sample

The geographical distribution of patients is depicted in Map 1. The largest proportion of patients (52,7%) were from the Cape Peninsula and vicinity, also the major catchment area of the hospital. As will be discussed later in the study, distance from the hospital posed its own problems to families who were further afield, both financially and due to lengthy separation. The map furthermore serves to indicate that the problems experienced by parents at both the maternity home and hospital, were not peculiar to a specific institution, but relevant to many hospitals throughout the Republic of South Africa.

For field work the very wide scatter of the rest of the sample resulted in travelling well over 8,000 kilometres in home visits and tracing.

For the purposes of the present analysis and within the limits of time and cost, the sample can be regarded as adequate.

GEOGRAPHICAL DISTRIBUTION OF PATIENTS IN THE SAMPLE



6. Retrospective study

Having secured the sample in the manner described earlier, a retrospective study was done of the 90 patients and their families. This type of cross-sectional study normally has two known disadvantages. Firstly it is static, with observations being made at one point in time and is therefore not geared to taking developmental aspects of interpersonal relationships into account. In other words it is, "....based on the assumption that the parent will continue to 'accept' the child, once he has done so, or that stress emanating from a 2-year-old handicapped child, will be perceived in a similar fashion when this child has reached adolescence" (Minde, et al.,1972 p.1554). Secondly the potential distorting influence of recalling past events is said to detract from the reliability of reports.

In order to counteract these limitations, an eclectic research design was followed:

(a) Systematic factfinding was done over a 2 year period. Thus the first full scheduled interview with mothers and/or fathers was succeeded by further home or office interviews and correspondence, in certain respects emulating the panel study approach, by repeatedly gaining information from the same sample.

(b) Additional sources of information viz. records of community based welfare organizations, general practitioners, hospital social workers, stomatherapists

and probation officers were tapped in an attempt to limit distortion.

(c) By studying families who had attended the hospital over a twenty year period i.e. 1961-1980, it was hoped to gain insight into both short and long term adaptations, as at that point in time and therefore less prone to distortion.

7. Interviews

Interviews were conducted by means of a semi-structured schedule comprising open-ended questions allowing free expression of feelings related to the experience of the birth of the patient, hospitalization and treatment and subsequent adjustment at home and in the community. Probes were used to gain indepth information and by necessity to direct responses to specific areas. Both home and hospital interviews were tape recorded and then typed on a word processor. With the aid of the latter each reference made to a specific topic throughout an entire interview was extracted and compiled separately. As researchers before have faced the paradox of denial to a direct question, later followed by spontaneous admission during the same interview (Jeffcote, Humphrey and Lloyd, 1979), this was a means of getting closer to the respondent's intention and also noting qualified responses.

In some ways the process followed here aided attempts at quantifying the material, a known difficulty when dealing with data which is largely qualitative in nature (Frydman,1979).

D. PROBLEMS EXPERIENCED IN THE STUDY

1. The main problems faced in exploratory studies are twofold: firstly, that of an overload of information and secondly, the difficulty with the quantification of data. The present study is no exception. Although every effort was made to present exact figures, the first priority remained to describe the C-factor i.e. the meaning attached to the birth of the child with an anomaly. For this reason the study draws heavily on direct citations from the interviews while quantification serves to indicate the proportion of parents projecting similar interpretations.

2. By the nature of the subject matter of the study, many of the findings are equally applicable to both the social work and medical professions. It was therefore impractical to seperate the recommendations into those pertaining purely to the role of the social worker on the one hand, and to that of the doctor on the other. This serves to further emphasize the fact that a team approach is essential in the management of the child with a congenital anomaly.

E. DEFINITION OF TERMS

By the nature of its subject matter, many terms in the present study are medical and essentially foreign to the social work profession. In order to facilitate clarity and understanding, such terms are grouped together and defined in a glossary (Annexure A) based on the Dorland's Medical Dictionary (1974).

The term "psychosocial" is frequently referred to in the present study. Although it is widely used in the literature pertaining to the family of the handicapped and chronically ill child, no clearcut definition could be found. For the purposes of the present study this term is used in its broadest sense, in essence as described by Hollis in the Encyclopedia of Social Work: "Psychosocial approaches are concerned with both the inner psychological realities of man and the social context in which he lives" (National Association of Social Workers, 1971, p.1217).

F. OVERVIEW OF THE STUDY

In the following two chapters the main findings in the literature pertaining to cystic fibrosis (Chapter 2), oesophageal atresia, Hirschsprung's disease and anorectal malformations (Chapter 3) are discussed. In order to meaningfully interpret the psychosocial aspects as found in the literature, these findings are preceded by a brief synopsis of the aetiology, clinical features and treatment of each anomaly.

The presentation of results of the present study broadly follows a chronological sequence of the difficulties experienced by the families of children with congenital anomalies, as from birth through school-going age. The findings related to the period of first awareness of symptoms to the time of learning the diagnosis of the child, is presented in Chapter 4. The confrontational phase i.e. coping with the acute stage of the birth defect is discussed in Chapter 5 followed by results on the difficulties experienced in the community (Chapter 6) and within the family itself (Chapter 7). The school experiences of patients are reviewed in Chapter 8, followed in Chapter 9 by a discussion of the findings pertaining to those families where patients had died.

In conclusion the recommendations based on the major findings of the study are presented in Chapter 10.

CHAPTER 2

THE CHILD WITH CYSTIC FIBROSIS AND HIS FAMILY: A LITERATURE STUDY

In the present Chapter the available literature on the psychosocial effects of congenital anomalies is reviewed. Research on cystic fibrosis (referred to in this Chapter as CF) is looked at first, followed in Chapter 3 by literature pertaining to oesophageal atresia, Hirschsprung's disease and anorectal malformations. In the same chapter some theoretical hypotheses of families under stress will be discussed, as found mainly in the sociological literature. In order to meaningfully interpret the findings of the various studies, each section will be preceded by a brief description of the specific anomaly and it's current treatment program.

A. CYSTIC FIBROSIS: THE DISEASE

CF is the commonest lethal genetic disease of childhood in Caucasians, yet till recently was unknown to many people. Being an inborn error of metabolism it involves pathological changes of the lungs and pancreas, and was therefore often mistaken for bronchitis, asthma or pneumonia. At the same time its digestive manifestations were frequently attributed to other illnesses of malabsorption. In 1938 CF was identified as a seperate clinical entity, but in the 1940's patients still rarely lived much beyond infancy. With the advent of antibiotics and improved diagnostic and treatment procedures, a longer life-expectancy as well as better health has become possible and today many children live to complete their school years. Seen predominantly in whites, one in

every 2000-2500 children born to this race group will be affected. The disease is transmitted by recessive inheritance and it is estimated that one in every 20 whites is a carrier. Only when both parents are carriers, is there a 1:4 chance of their producing an affected child. CF is neither detectable by amniocentesis, nor can carriers be identified by any test.

Alternatively called mucoviscidosis, this disorder affects the exocrine glands resulting in the production of abnormally thick and sticky mucus, causing blockage of some of the internal organs. In the lungs this leads to recurrent infections as patients have extreme difficulty in coughing up the viscid mucus, which then forms a breeding ground for bacteria. The eventual lung damage leads to respiratory problems which again gradually compromises heart function. In the pancreas and liver, thickened mucus interferes with the normal production and flow of enzymes and the secretion of bile salts, with the result that fat, proteins and fat-soluble vitamins are not well digested or stored by the body, giving rise to malabsorption with bulky, foul smelling stools in a thin and malnourished patient.

The treatment of CF is predominantly geared to the prevention of chest infections by keeping the lungs clear. The time-consuming regimen at home includes daily to 3 times daily inhalations in a mist tent, which helps to liquify the mucus, making it easier to cough up. In this the patient is further assisted by vigorous physiotherapy and suctioning by machine. A room humidifier is often run at night to ease breathing and facilitate gaseous exchange in the lungs. In addition most patients are maintained

prophylactically on antibiotics which frequently require changing when infections nevertheless develop. Severe infections may require hospital treatment.

Pancreatic involvement is managed by giving pancreatic enzymes with every meal, resulting in an increased absorption of protein and fat in the patient. Supplementary vitamins are also administered daily.

No cure exists for CF as yet, and it is important that treatment start as early as possible before permanent damage is done. The still frequent difficulty of obtaining a positive diagnosis, remains a major problem. Although a relatively uncomplicated sweat test, revealing an abnormally high secretion of sodium chloride in CF patients, serves to confirm the diagnosis, the initial symptoms are often misinterpreted. Once diagnosed, parents should realise that with constant adherence to the treatment regimen, periods of relatively good health can be experienced. The prognosis is uncertain, depending on many factors, inter alia severity of the illness, the time of diagnosis, cooperation of the patient, as well as family and medical management.

B. PSYCHOSOCIAL ASPECTS OF CYSTIC FIBROSIS

In the discussion of psycho-social aspects of CF (Gayton,1977, Frydman, 1979; Lewis, 1981) frequent reference is made to the "early studies", (Turk, 1964; Kulzcycki, 1969; McCollum, 1970; Tropauer, 1970) where emotional distress, economic pressures and problems in family functioning had been consistent findings. The

following authors were of opinion that the nature of these results could be questioned due to methodological shortcomings such as a lack of comparison groups (Drotar et al, 1981), making use of subjective evaluations (Lewis and Khaw, 1982), being based on clinical impressions (Gayton et al., 1977) and being derived from small or biased samples (Frydman, 1979).

In contrast to the above the more recent research (Bywater,1981; Straker, 1980; Mittenzwey et al., 1981; and Lewis, 1982) predominantly projects positive conclusions, with little significant difference due to CF found in the factors studied by them.

In a further handful of studies the discrepancy in overall findings between the two groups mentioned above, is bridged with a conclusive view which is distinctly moderate. Three separate studies (Burton, 1975; Falkman, 1977 and Tavormina, 1981) for instance report normal to poor functioning in certain respects while reflecting a positive view on others.

Due to the very wide range of methods used, the differences in age groups studied and the specific aspects looked at, it has remained a problem to scientifically compare results or challenge previous findings. The present reasoning was that results could only be meaningful when read within the context of the sample population, background and objectives of the particular study. Therefore to facilitate further discussion, a synopsis of the studies representative of the three mainstreams of thought identified above, is presented in schedule 1. In this way an attempt is made

SCHEDULE 1 GROUP A

PSYCO-SOCIAL ASPECTS OF CYSTIC FIBROSIS: A SYNOPSIS OF RESEARCH STUDIES

AUTHOR	ORIGIN	SAMPLE SIZE	AGE GROUP	M E T H O D S	RESPONDENTS	TOPIC	SAMPLE SELECTION	POSSIBLE BIAS
1.Turk (1964)	Maryland USA	25 families 53 patients	3m-23yrs	Questionnaire: Forced choice & open end	Mothers-25 Fathers-3	Deprivation due to CF	All pts.attending clinic in 67/52 period	
2.Kulczycki et al (1969)	Washington DC USA	20 families 26 patients	10 yrs	"Psychiatric & psychological evaluations" "Standard tests of intelligence"	Parents & patients	Effect of CF on pt Parent response	Not described	
3.McCollum (1970)	Yale Connecticut USA	56 families 65 patients	0- 10yrs	Multiple choice Questionnaire Interviews Group Discussions	Parents	Adaptive stages of family to CF identified	Not described	
4.Trophauer et al (1970)	Dayton Ohio USA	23 mothers 20 patients	5-20yrs	Psychiatric Interviews	Mothers & patients	Psychological aspects of caring for CF child	Selected on basis of availability for psychological examination	
5.McCollum (1971)	Yale, Connecticut, USA	54 families 62 patients	Unknown	Cost of care followed for one year	Parents	Cost	All families attending centre for full year	
6.McCrae et al (1973)	Edinburgh Scotland	50 Scotland 50 N-Ireland families	Unknown	Int.on intellectual awareness emotional & behavioural response to genetics of CF	99 Mothers 87 Fathers	Genetics	Not described	

GROUP A (cont.)

AUTHOR	ORIGIN	SAMPLE SIZE	AGE GROUP	M E T H O D S	RESPONDENTS	MAIN TOPIC	SAMPLE SELECTION	POSSIBLE BIAS
7.Allan et al (1974)	Melbourne Australia	50 Mothers of CF pts.	0-29	Interview	Mothers	Family response to CF	"Sampled from 174 public & private pts at Royal Child Hospital"	Excluded where pt. was terminal Mother under psych treatment.Death or diag.of CF less than 1 year before int.
8.Boyle et al (1976)	Maryland Washington,DC	25 Families 27 Patients	13-30yrs	Psych.int. W.A.I.S. W.I.S.C. Thematic Apperception Test; Rorschach Test D-a-P Test	Patients 21 Mothers 4 Fathers	Emotional adjustment of young people with CF	Not described	
9.Mikkelsen et al (1978)	California USA	18 Families 19 Patients	4-11yrs	Interview	Patients Parents	Living with CF: A family challenge	Not described	Approx 1/3 of sample hospitalized at time of int.Min. qualification.
10.Kerner et al (1979)	Palo Alto, Calif. USA	16 Families who had lost a child with CF	6-24yrs	Interview	Parents	Grief	Parents of all pts who died at Stanford Hosp. 1971-1975	4 families could not be traced.

GROUP B

AUTHOR	ORIGIN	SAMPLE SIZE	AGE GROUP	M E T H O D S	RESPONDENTS	MAIN TOPIC	SAMPLE SELECTION	POSSIBLE BIAS
1.Wagner et al (1976)	Rostock Germany	21 Parents of patients	4-16yrs	Standardized interviews Standard Intelligence Tests	Parents	Parents reaction to diag,school & family functioning	Not described	Half of pts were so well that treatment regimen not adhered to
2.Gayton et al (1977)	New York U.S.A.	43 families	5-18 yrs	Semistructured interview schedules Parents: Family-concept Q sort M.M.P.I. Patients: Piers-Harris Self-Concept Scale Missouri Children's Picture Series Holtzman Inkblot Test	Parents Patients Siblings	Psychological test findings of CF pts,sibs & parents	43 of 72 pts on masterlist except those names under "exclusions"	11 moved, could not be traced; 5 indirect refusals 4 pts were adults - excluded 2 pts died 4 refused participation.
3.Straker & Kuttner (1980)	Jo'burg R.S.A.	10 CF pts 10 healthy controls	12-16 yrs	Clinical int. Norwicki-Strickland Locus of Cont. Test;Thematic Apt. Test	Patients & controls	Adolescents with CF:Psychological dimensions	Matched control group study	--
4.Mittenzwey (1981)	Dresden	260 parents of CF pts.	Unknown	Postal Questionnaire	Parents of CF pts	Psycho-soc problems of families with CF	Population unknown	Return rate unknown

GROUP B (cont.)

AUTHOR	ORIGIN	SAMPLE SIZE	AGE GROUP	M E T H O D S	RESPONDENTS	MAIN TOPIC	SAMPLE SELECTION	POSSIBLE BIAS
5.Drotar et al (1981)	Cleveland Ohio,USA	91 CF 47 Chronic illness 71 sibs of of CF pts 61 healthy children	3-13 yrs	Parents:Mailed checklists Louisville Behaviour checklist Teachers: School Behaviour Checklist.	Parents Teachers	Adjustment of CF children compared to other groups	All pts of those diagnosis receiving care at specific centres were mailed check lists	Return rate 67%-55%
6.Bywater (1981)	London,UK	27 patients	12-16 yrs	Mothers: Semi-structured interview Malaise Invent. Teachers: Rutter Child Scale B2 Patient: Semi-structured interview	Mothers Teachers Patients	Psychological adjust of adolesc. with CF	All pts (30) between 12-16 invited to take part	3/30 did not agree to cooperate
7.Lewis and Khan (1982)	Boston Mass. USA	31 CF 26 asthmat 27 healthy	7-12 yrs	Mothers: Family Adaptability and Cohesiveness & Evaluation Scales Behaviour Problem Checklist Children: Piers-Harris Children's Self-Concept Scale	Mothers Children	Family functioning affecting adjustment of CF child	Randomly selected from treatment population	Excluded (i)if there affected sibs (ii)if pt had been hospitalized the preceding year

GROUP C

AUTHOR	ORIGIN	SAMPLE SIZE	AGE GROUP	METHODS	RESPONDENTS	T O P I C	SAMPLE SELECTION	POSSIBLE BIAS
1.Burton (1975)	N-Ireland	53 families 58 patients	7/12-16yrs	Parents:Interview Patients:Vineland Soc Mat Scale Draw-a-man Schonell Word Read Test Schonell Spel Test Vernon Arithmetic/ Maths Test Taylor Manifest Anxiety Scale T.A.T. Teachers:Bristol Soc.Adjust.Guide	Parents Patients	The family life of CF children	54 families who could be traced from all known families in N-Ireland	No of untraceable patients not mentioned
2.Falkman (1977)	Sweden	52 patients	4½ - schoolage	Patients:Leiter Int Perform Scale W.I.S.C. Bender Visual Motor Gestalt Test Human Fig Draw Mothers:Interview E.P.I. Teachers: Questionnaire	Mothers Patients	Special characteristics of CF children & their families	Absolutely true random sample	No missing cases Good sampling
3.Tavormina (1981)	Illinois USA	144 families of CF,diabetic,asthmatic & hearing impaired patients	5-19 yrs	Hereford Parent Att Survey E.P.I. Missouri Behaviour Problem Checklist Special Problem Area Check List Vander Veen Fam Unit Inventory	133 mothers 93 fathers	Coping strategies of parents with a handicapped child	Not described	16 families did not consent

at obtaining perspective as the different authors are cited.

Turk, the first author in Group A of Schedule 1, is the most frequently quoted as the first of the earlier studies which set out to determine whether the time and cost required in caring for a CF child, resulted in other members of the family being socially or financially deprived. Although basic needs like food and shelter were not in jeopardy, the families in this study felt that adequate time and means for adult activities, time for self and for leisure was not available due to the constraints of CF in these areas. A serious added problem was the resulting lack of communication between family members which is referred to as "the web of silence", which in turn caused misunderstanding and impaired family functioning.

In the years following, these findings have been either wholly or partly supported and often challenged as more controlled research designs addressed more specific issues in this respect. The main issues thus raised and the arguments for and against will form the contents of the following discussion.

1. Economic impact of cystic fibrosis on the family

Hospital and clinic treatment of CF is free of charge in most of the major treatment centres of the world. Yet a number of studies especially from the USA and Australia were in agreement that in spite of this, finance was one of the areas highly charged with stress. The families in Turk's study (1964), although claiming not to be deprived of the essentials of living, reported a significant breakdown in communication between spouses on this topic as well as

having to forfeit family holidays in order to make ends meet.

In 1971, McCollum carefully examined both the overt and hidden costs of children in the Yale Cystic Fibrosis Program retrospectively over a one-year period. Of the families studied, 79% were in supplementary employment in order to meet the extra financial commitments of CF. Again vacations away from home were only possible once in many years while the actual cost to the family for the care of the CF child alone, had reached the alarming proportion of almost one-quarter of their income. She concludes with special concern for those families who had fully extended their earning capacity in attaining a moderate income, yet were reduced to extreme financial hardship by this chronic illness.

As was expected, the same extent of financial burden was not present in a welfare state, such as Northern Ireland (Burton 1975). However costs which did affect some families were amongst others, transport to hospitals and clinics (73%), special diets (45%), and disrupted work patterns with the added loss of pay (11%).

Considerable differences in cost were found in 50 Victorian families (Allan et all 1974), with 30% never having experienced financial problems, while 24% reported on-going and severe financial difficulties. The largest proportion (46%) had had problems in the past, which is best explained by the fact that the period of greatest health expense was found to be before the diagnosis was established and immediately thereafter when expensive equipment such as mist tents and humidifiers had to be purchased. A source of additional expense which although not medically prescribed but was nevertheless found to be essential in other

studies (McCollum 1970) to counteract the effect of mist tents, was the purchase of extra heaters and air cooling equipment. Such attempts at facilitating home treatment, together with less costly but nevertheless continuously needed items like extra toilet cleansers, room deodorants and special foods, were covert costs which families had to accommodate within their normal budget, as were buying clothes of more expensive natural fibre rather than synthetics which would further aggravate the excessive sweating of CF patients. The above researchers were in agreement that leisure was the budget item most often omitted in favour of these necessities.

2. Marital relationships in families with cystic fibrosis

Many studies report instances of separation and divorce in families caring for a CF child, but samples are small and rarely compared to the national figures of the countries of origin. Of the 26 families studied by Kulczycki (1969), 3 fathers had left; 9 of the 56 couples in McCollums's (1970) sample were divorced or separated; Gayton (1977) mentions that 5 of the 43 fathers were not seen because of divorce and McCrae (1973) found 8 separations and 1 divorce in his 50 Scotland families as opposed to only 2 separations in an equal number of North-Ireland families.

In an attempt to meaningfully analyze the various sources quoting incidence of divorce and separation in couples with chronically ill children, Begleiter et al (1976) used the data of 14 other researchers and compared the pooled results with their own and the statistics for the general population of the United States of

America. Admitting to the limitations of including populations outside the USA, the overall finding was nevertheless that prevalence of divorce and separation was no higher for parents of children with chronic diseases, including CF, than in the general population. The CF couples alone had a higher incidence than the other chronic disorders, i.e. spina bifida and leukemia, for which the rate was below the national average. The authors reasoned that a possible explanation for the latter discrepancy was the greater genetic risk attached to planning further pregnancies for the CF families.

In the very soundly constructed study of Falkman (1977), based on a true random sample of all CF families in Sweden, 4 of the 50 couples were divorced, 3 directly blaming CF. This was slightly below the national mean and, the author speculated, probably due to the excellent financial support of the Swedish Health Insurance to these families. However 26% of her sample felt that their marital relationship had deteriorated. The majority of this group were also reporting an inability to communicate about CF, a factor which was also noted by Kulczycki (1969) and Turk (1964). Falkman detected over-involvement of the mothers with patients in this group, reducing the father to the periphery in family relationships. In an Australian sample (Allan et al, 1974), an observation of similar nature was made: one quarter of the marriages had bonded more closely as a result of having a CF child, mainly where fathers had been actively involved in care of the patient and support of the mother in time of stress. The same sample had 33% discordant marriages of which 6% were divorced and 14% confessed to complete breakdown, but living at home for the

sake of the patient.

Optimistic findings of unchanged or even closer marital relationships in CF families are found in the literature (Wagner 1976). However interpretations need to be made with great caution. Closer scrutiny of Schedule 1 reveals that samples are often biased towards inclusion of stable families i.e. those who could be traced, those who attended clinics regularly, those who agreed to participate or those available for testing. Furthermore, the absence of divorce cannot unequivocally be seen to represent marital stability. In Burton's study (1975) 50% of the women felt that the marital relationship was strained, 20% felt sexual relationships had been destroyed, yet no divorce was mentioned in the findings. Factors militating against stable marriages, like lack of money for leisure, lack of communication, the risk involved in further pregnancies, limited time due to lengthy treatment regimens and curbed holiday possibilities were certainly present in research findings.

3. Parental response to cystic fibrosis

(a) The pre-diagnostic stage

Parental response to CF was seen by McCollum and Gibson (1970) as developing through four major stages. The time between onset of parental concern for the patient's health and establishing the diagnosis of CF, was termed the pre-diagnostic stage, which in their sample had a mean duration of 17½ months and a maximum of 8 years. This stage was characterized by repeated fruitless attempts at obtaining a diagnosis, accompanied by rising anger and hostility

directed at the medical profession when diagnoses like feeding problems, allergic reaction or parental overconcern were made. This dilemma is confirmed by Mikkelsen et al (1978) where 50% of the sample had a similar problem; Falkman (1977) where 71% of the diagnoses were made between 3 months and 7 years after parents' initial concern, and Burton (1975) where 40% were given a faulty diagnosis - 23% mothers being told that they were fussy. The main impressions associated with this stage by other researchers were: considerable financial expense (Allan et al 1974); the striking initiative in the efforts of parents to obtain a correct diagnosis (Falkman (1977) and a harmful effect on the mother-child relationship (McCollum and Gibson 1970). The latter project also noted anxiety and the increasing loss of confidence in mothers concerning their maternal abilities, often followed by depression.

(b) The confrontational stage

McCollum and Gibson (1970) furthermore describe the confrontational stage when the diagnosis was confirmed. This stage is distinguished by initial denial, followed by anticipatory mourning, a phenomenon also described by others (Solnit and Stark, 1961). Fear of losing the patient was seen as a central issue, with associated problems of subsequent pregnancies to replace the patient, yet fearing the genetic risk involved. At the same time anger was directed at physicians for medical indecision, at the marital partner for the genetic inheritance and towards God and the church in general. Overt anger towards the patient was however, seldom seen at this stage. Kulczycki et al (1969) contrast by finding some ambivalance and rejection towards the patient, albeit transient, with occasional death wishes for the child.

Paradoxically the majority of parents in their study also expressed relief at the time of diagnosis, as ending many uncertainties. Kerner et al (1979) in their study of 16, families found only one couple rejecting religion, and 5 families seeing it as a major source of support. An unopposed view was the initial inability of parents at this stage to fully grasp the meaning and implications of information given to them. This may well explain the disappointing findings where knowledge of the genetic base of CF was measured at a later stage. At this time parents were embroiled in altering the physical environment to the patient's needs, mastering therapeutic techniques and dealing with the reaction of siblings. From the German literature, Wagner et al (1976) felt that their mothers reacted more adequately than their American counterparts, in that their overall attitude did not indicate exaggerated response, i.e. they hardly changed their work or activities and most patients attended nursery schools. Some explanation for this discrepancy may be found in the fact that half of these patients were so well that therapy was not even adhered to. (See Schedule 1, group B).

(c) The long-term adaptational stage

The foremost demand posed by the long-term adaptation stage, was to sustain a mutually satisfying relationship between parents and a potentially dying child. Parents seemed to vacillate between denying CF, and mourning the impending death of their child. Denial as a coping mechanism became difficult in the face of the pervasive CF odour, the typical sounds of humidifiers and the hacking CF cough. An average of 1.6 hours per day was spent solely

on therapy and encroached on and interrupted other family activities. The time factor was also raised by Turk (1964), and in his study of 260 families, Mittenzwey (1981) found that care of affected children required a minimum of 2 - 3 hours a day, with therapy being mainly performed by the mothers. Many other studies reflect the fact that the bulk of the burden of CF therapy fell on the mother. Of the 52 families studied by Falkman (1977), only 6 fathers gave daily help and 37% had never participated in therapy. In the sample of McCollum and Gibson (1970), 57% fathers of the intact families helped their wives, while 43% rarely, if ever, did so. Oppenheimer and Rucker (1980) report 43% households sharing treatment, while Boyle et al (1976) pleads for greater involvement of the father, and Tavormina et al (1981) refers to the 'emotional divorce' of the father in this respect. One contributing factor mentioned was that the diagnosis was often given to the mother alone (Falkman,1977). Among the otherwise optimistic and positive findings in her study of 27 adolescents with CF, compared with a group of healthy adolescents and their mothers, the only negative finding of Bywater (1981), was the significantly higher incidence of depression in mothers of CF patients. Her recommendation that 'special help should be given to the mothers as they appeared to carry the main burden of coping with the illness', echoes the findings of the researchers mentioned above and is fully supported by Stephan and Biener (1978). A further study where depression in mothers of CF patients was measured by means of a formal scale (see Schedule 1), confirms this raised incidence (Gayton, et al 1977), while reported depression was found in 79% of the mothers in the sample of Mc Crae, et al (1973), 42% of them seeking medical help in this respect. Some mothers resolved by finding work outside the

home, mostly part-time, primarily to avoid being cooped up with CF problems for a full day. In Bywater's (1981) sample, 67% mothers worked, including two-thirds of the most severely affected cases. Boyle et al (1976) found this a mutually beneficial arrangement, as it tended to counteract infantilization of the patient: of their patients doing well, 70% of mothers were gainfully employed. The oft repeated finding of mothers feeling isolated by CF, seemed also to have been relieved by their entering the work situation.

Findings on the long-term adaptation of fathers to CF are less clear. Their reactions have been inadequately studied, mainly because they have not been available for testing, and researchers have also seen the mother as the central determinant of the CF patient's well-being. Perusal of research findings therefore shows with almost monotonous regularity that the opinion of the father was either proportionately under-represented or not accounted for at all (see Schedule 1). Depression however, seems not to have been a significant problem (Bywater, 1981; Gayton et al,1977). Where formal personality testing was performed by the last mentioned researchers, 32% of the fathers and 22% of mothers scored within ranks suggestive of emotional disturbance. They conclude that living with a CF child was more likely to result in impaired personality functioning for the father than the mother and offer as a possible explanation the extreme pressure on the breadwinner due to the economic impact of the illness. Their final view was that the extent of deviation recorded was not as severe as other studies indicated and that the principal effect of CF was more prevalent in a lowered adjustment and satisfaction of the family as a whole. This seems to complement the findings of Cummings (1976), who

assessed 240 fathers of healthy, retarded and chronically ill children including CF, where the third group mainly demonstrated significantly decreased states of gratification in interpersonal relationships. To a lesser extent self-esteem - notably the component referring to paternal adequacy - was unfavourably influenced by a chronically ill child. When comparing these results on fathers to earlier results on mothers (Cummings et al, 1966), more frequent indicators of stress reach statistical significance in the personality variables of fathers than mothers. This is explained in part by the greater involvement of mothers in therapy, which the authors reason, counteracts feelings of helplessness and guilt.

In contrast Tropauer et al (1970) noticed that emotional upset in mothers coincided with setbacks in the patient's health. Mothers questioned whether they could not have done more and were inclined to overestimate the potential effect of their efforts. This was due to the relative importance normally attached to involving parents in active treatment of CF. The authors argued that, far from expending guilt and depression by their treatment efforts, these feelings could be intensified.

Tavormina et al (1981) who studied coping behaviour of both fathers and mothers of children with congenital anomalies, including CF, report not only more frequent coping problems in mothers, but also more detrimental effects due to CF than found in fathers. As a group the parents projected a need to be accepted as 'good parents', at the same time doubting their own parental skills. The overall finding of this project accentuates the fundamental

normality of the group, but as reacting to the extreme demands posed by the realities of raising chronically ill children. Observed symptoms were therefore not of personally internal origin, but from the reality issues faced by parents. Even in the face of these problems, other researchers mention the tendency of parents to minimize the problems of CF when interviewed for research purposes (Burton 1975).Further aspects of the long term adaptational stage reported by McCollum and Gibson (1970), are specifically related to the age or stage of development of the CF patient. For the purposes of the present study, these will be discussed with the findings relating to the patients' adjustment to CF. Some aspects of the terminal stage, which was mentioned but not discussed by McCollum and Gibson (1970), will be looked at in conclusion.

4.Family functioning and CF

In the discussion so far, aspects of family functioning was referred to intermittently. It has become clear that families did have restrictions in time as well as communication and financial means, which could influence family functioning negatively. Some extent of marital and family strain due to CF was also noted in the literature discussed earlier. The question arises how well the CF patient could adjust where such impaired family functioning was present.

Gayton et al (1977) and Tavormina et al (1981) who made use of standardized tests, found strong agreement on the assessments of mothers and fathers on most factors regarding family functioning.

Of importance here are the findings of the latter project, firstly, that parents saw their life as centered around the CF patient and secondly, their significantly higher than normal scores on the cause factor, which was described as 'the extent to which the parent sees himself as a major factor in determining the child's behaviour.' (Tavormina et al, 1981, p.123). Similarly, the Family-Concept Q Sort test results of Gayton et al (1977) revealed that CF parents saw potential for better family satisfaction and family adjustment with a non CF child than in their own situation at the time of testing. The reciprocal nature of family functioning and caring for a CF child is further endorsed by Oppenheimer and Rucker (1980). Their study of 37 families confirmed that more effective home treatment was given to those patients living at home with both their biological parents than the patients where family breakdown had occurred. This was also reflected in the better physical condition of the patients in the former group.

Employing a number of tests (See Schedule 1, Group B), Lewis and Khaw (1982) compared the self-concept and frequency of behaviour problems in chronically ill children, including CF, with a group of healthy children. The results showed family functioning to be a better predictor of child adjustment than the presence or absence of disease. Family functioning was therefore isolated as an important mediating variable in affecting psycho-social adjustment in children with CF. The authors further hypothesized that impaired family functioning need not be of permanent nature, but could be limited to particular episodes of stressful demands, after which time, movement back towards healthy functioning should again occur. Truly dysfunctional families can then be identified by the

inability to move back to sound functioning once a crisis has been resolved.

5. Genetic issues in CF

In view of the considerable stress suffered by families caring for a CF child, genetic counseling assumes crucial importance. Yet studies from the UK (Burton, 1975; McCrae et al 1973), Australia (Allan et al 1974), Sweden (Falkman, 1977) and the USA (Kerner et al 1978), report subsequent pregnancies, which were both wanted and unwanted.

In the Swedish study mentioned above, parents who had a healthy child or children before the affected child was born, were less likely to have further children than where the CF child was a first-born, in both instances following correct genetic information. The strong desire for at least one healthy child, together with widely differing interpretations of the odds of repetition, was felt to be of major importance. Religion seemed to be the factor accountable for further pregnancies in the sample of Kerner (1978), where 12 of the 16 couples had had genetic counseling. The majority (75%) then decided against further pregnancies, while the 25% who continued expanding their families, were all Roman Catholics.

Where comprehension of genetics was looked at in the two UK studies, the results were poor. In both samples a sound grasp of the genetic base of CF was limited to 20-22% of parents. In Burton's study (1975), mothers were proportionally more

knowledgeable than fathers. However, when analyzed by social class, fathers in classes I and II ranked best as a group, which was partly explained by the fact that they were also the group who had made the most effort on their own to gain more information on the illness. McCrae et al (1973) confirm the foregoing, but add that the principal causes in their sample were psychological barriers to genetic information as well as badly timed counselling by doctors. If parents originally had no conflicts about limiting their families after the CF child was born, contraception was more successfully practised. Otherwise decisions were based mainly on subjective factors like the demands made by the care of the CF child. This is confirmed by Leonard et al (1972), who studied the reproductive dispositions of 76 families caring for children with phenylketonuria, Down's syndrome and CF. Although the families in this carefully designed study had all been subject to genetic counselling, the authors conclude that, "... reproductive attitudes are determined more by the sense of burden imparted by the disease than by knowledge of its precise risk figures" (p.433). In spite of counselling, only half of the sample had gained a good understanding of the information given to them. The explanations given by the authors include: lack of knowledge of human biology; variable skills in counselling as well as parents' views on the usefulness of genetic counselling. In a further analysis however, no difference in knowledge could be found between the group who continued with subsequent pregnancies and those who did not. Later in the study religion is mentioned as a major impediment to the implementation of such knowledge.

In all studies the greater majority of parents, roughly 70-75%,

were in favour of no further pregnancies. However, this intention was seldom enough to ensure successful family planning. Continued genetic and contraceptive advice repeated at different stages as advocated by McCrae et al (1973),is probably indicated, as parents from some studies pointed out that inadequate information had been given about the illness (Falkman, 1977; Burton, 1975).

No clearcut findings on the emotional response to the genetic base of CF could be found. Whereas McCollum and Schwartz(1972) describe anger towards the marital partner where a disease is genetically transmitted, a direct question to parents in the study of McCrae et al (1973) revealed such resentment and blame to be uncommon.

6. Impact on siblings of CF patients

The adjustment of healthy siblings has only been succintly dealt with in most of the literature pertaining to the affects of CF. In younger siblings parents have reported somatic complaints, pretending to have CF (Tropauer et al 1970) and viewing treatment given to the patient in the light of favouritism (Mikkelsen et al 1978). Older siblings were described as concerned and helpful (Mikkelsen et al 1978); as rendering protective care to the patient (Burton 1975) while the sibling next to the patient was seen as most lacking the mother's attention (Allan et al 1974). In the latter project one-third of siblings were reported to have behavioural problems.

Where a CF patient had died, Kerner et al (1979) found undue guilt in siblings to be rare. Although practically all of them confessed to having felt somewhat neglected, the response was at the same

time qualified by the fact that they had understood the reason. Teenage girls in this sample verbalized some concern about the possibility of being a CF carrier.

In a number of studies the findings were predominantly positive, for instance in the sample of Falkman (1977) stress often associated with the negative prognosis of CF was not found to be present at all in the siblings studied. Furthermore one of the first studies on sibling adjustment to make use of standardized tests in this respect Gayton et al (1977), found the scores well within normal limits, offering little support for detrimental psychological impact on siblings.

Breslau et al (1981) devised a study to specifically measure psychological impairment in healthy siblings of sick children compared to 1034 randomly selected children. Siblings of 239 children with CF, cerebral palsy, myelodysplasia and multiple handicaps were studied in this way. The overall result did not verify earlier impressions of significantly higher incidence of serious malfunctioning in the studied children. However, on certain sub-scales measuring aggression towards peers and at school, significantly greater pathology was indicated. Nevertheless it was explicitly stated that this could not be linked in any way as a response to parental pre-occupation with the patient. This argument was based on the finding that there was no association between the adjustment of the sibling and the level of disability of the patient. In contrast to the authors cited earlier, the younger siblings were not worse off than the older ones. On the other hand female siblings older than the patient,

were found to be at risk, which they suggest should be empirically researched before explanations are offered. Rather than seeking association between sibling pathology and mother's attention, the authors recommend that the two following factors be investigated: the effect of the mother's strained mental and physical health on siblings, and the direct influence of patients on siblings in the spheres of body image and learned behaviour.

7.The impact of CF on the patient

Chronic illness brings different challenges and difficulties at each stage of development for the CF child. McCollum and Gibson (1970) described the first year of life and beyond as manifesting feeding problems originating from an insatiable appetite. Normal developmental issues like, inter alia bowel training, were aggravated by the increased frequency and foul odour of stools while exposure to new experiences were curbed for fear of infection. The necessary independence had not been reached at school age and 44% of patients manifested adjustment problems during the first year of attendance. The importance of peer acceptance became a discomfort between 8-12 years, due to symptoms like tiredness, flatulence and treatment regimens which were seen as obstacles to conformance to the peer group. A growing conciousness of the terminal nature of the illness was often amplified by insensitive reference by pupils and siblings to the patients' imminent death. During the teens delayed physical development was accentuated, accompanied by a mounting resistance to therapy, to which parents acquiesced, prompted by feelings of futility.

Other researchers have studied the impact of CF mostly during adolescence on specific aspects of patient functioning, their IQ and school adjustment. In the more recent studies, overall findings have been more optimistic, as will become clear from the rest of the discussion, which is presented more or less chronologically. This optimism was partly due to more strict research design, as well as the improved quality of life which has become possible for CF children.

By administering an abbreviated form of the House-Tree-Person technique to 20 children with CF, Tropauer et al (1970) reported two or more indicators of emotional disturbance in at least 70% of their sample. It was found that in 75% of cases feelings of inadequacy and/or insecurity was mirrored in the drawings and that patients' anxiety was related to the image of themselves, but not exclusively so. The authors reasoned that, although discoverable, anxiety had not reached the level of influencing observable behaviour, which accounted for the discrepancy between patients' concerns and mothers' accounts of their adjustment. During subsequent interviews, it was uncovered that worry about death, the future and restrictions due to CF added greatly to patient distress.

Six years later some of these findings were confirmed by Boyle et al (1976) who made use of a more extensive battery of tests. The four major areas of stress isolated in this study of 27 adults and young children, were firstly a distorted body image originating in their different physical appearance: all indicated dissatisfaction with their bodies, either due to delayed secondary sexual

characteristics or symptoms like excessive thinness. Related to this, was secondly a sense of isolation in most of the patients, emanating from a feeling of non-acceptance by the peer group. This seemed to be intensified by the third source of stress: parental conflict in upbringing which was mainly evident in families where discussion of CF was discouraged and the patient infantilized. On projective testing such patients displayed marked hostility towards parents and themselves. Finally, the authors identified an increased awareness of the future and death which was handled mainly by denial and refusal to talk about these issues. No relationship could be found between patient IQ and emotional coping abilities.

In the same year Wagner and Hein (1976) report that the depressive mood reflected by earlier studies, as well as the raised level of anxiety in CF patients, was not present in their sample. The latter was described as extroverted, happy and content with a negligible incidence of enuresis and sleep disturbance. Gayton and his associates, a year later (1977) continued in similar vein: in their sample prevalence of emotional disturbance was not disimilar from the normal population in both CF patients and their siblings. Earlier in the chapter (see Schedule 1, group B) areas of possible bias were pointed out in both the samples of Wagner and that of Gayton.This bias may well have influenced the positive outcomes reported above.

Falkman (1977), after careful review of the incidence of enuresis and sleep disturbance in the normal population, found a definitely raised incidence in her absolute random sample of CF children.

Furthermore, results on the Bender test reflected perceptual-motor disturbances, a finding supported by the emotional disturbances revealed by the Human Figure Drawings tests. In spite of a large proportion with reading and writing difficulties, the children in this sample also coped surprisingly well at school.

The latter finding was confirmed by three recent reseach projects. Bywater (1981), although noting a tendency towards depression, could not isolate any significant problems at school, with good communication in the families and denial of the eventual consequence of the illness contributing towards this positive finding. Drotar et al (1981) who studied four groups, including CF, indicated age-appropriate adjustment for CF patients at school with an absence of undue anxiety or depression. A possible explanation offered by the authors for the contrasting absence of these symptoms, was that their study did not evaluate patients while hospitalized - a period more likely to reveal higher levels of anxiety and depression. The third project (Mittenzwey et al,1981) related "exceptionally good" school adjustment, based on the questionnaires returned by 260 families with CF children.

Most studies record normal IQ's for CF children (Burton, 1975;Wagner and Hein, 1976; Falkman, 1977) and did not support Boyle et al (1976) who recorded above average intellectual functioning in his sample. Although patients in Burton's (1975) sample were well within normal limits of intelligence, 55% girls and 66% boys were retarded in two or more subjects at school. After careful contemplation of relevant factors, she concluded that the overall level of anxiety (as measured by the Taylor Manifest

Anxiety Scales) and patients' general response to school, were mediating variables in this respect.

A further contribution to the literature, emphasizing the normal psychological functioning of CF patients, was made by Straker and Kuttner (1980) who compared test results on the degree of control of a sample of 10 CF adolescents from Cape Town and Johannesburg with normal controls. (See Schedule 1, group B). Only one dimension, i.e. persecution, yielded a higher than normal score. The authors suggest that research on the development of coping mechanisms in CF patients be persued.

The majority of "optimistic" studies discussed were in agreement that patients were subject to major stresses emanating from CF as a chronic illness. In spite of this, many children managed to function on a level which did not reflect observable dysfunction. Some mediating variables acting contrary to coping mechanisms were identified. The central message seems to be that patients' strengths and coping mechanisms need to be supported and developed in order to prevent dysfunction.

8. Mourning the death of a child with cystic fibrosis

Of the authors studied, Kerner et al (1978) were the only to specifically address the impact of the CF patient's death on the family. Sixteen families were studied retrospectively, on average 2½ years after the patient had died. In seven of these families mourning was regarded as incomplete, either because the patient's room had been maintained as a shrine or weekly grave visits had

been continued for more than six months. Siblings in this sample had less problems than parents who showed a "high incidence of emotional and medical problems" (p.221). The authors suggest that counselling be offered to parents to aid with completion of the mourning process.

C. SUMMARY

In this chapter a brief outline of the clinical features of cystic fibrosis was given. Following this, the research literature on the psycho-social aspects of cystic fibrosis was reviewed. The findings indicate that families and patients were subject to major stresses due to cystic fibrosis. Yet many patients and families managed to function without observable dysfunction. However, indications were that families' coping mechanisms had been severely taxed and needed to be enhanced and developed to prevent the hardships accompanying cystic fibrosis.

In Chapter 3, a similar discussion of oesophageal atresia, Hirschsprung's disease and anorectal malformations is presented.

CHAPTER 3

THE CHILD WITH A CONGENITAL ANOMALY AND HIS FAMILY: A LITERATURE STUDY OF OESOPHAGEAL ATRESIA, HIRSCHSPRUNG'S DISEASE AND ANORECTAL MALFORMATIONS.

In the present Chapter the clinical features, treatment and prognosis of oesophageal atresia, Hirschsprung's disease and anorectal malformations are briefly outlined. This is individually followed by discussion of the available research literature on the psychosocial aspects pertaining to each anomaly. Because of the paucity of literature especially with regard to Hirschsprung's disease and anorectal malformations, some findings on handicap in general are discussed in conclusion.

A. OESOPHAGEAL ATRESIA: THE ANOMALY

Oesophageal atresia, shortly defined as a congenital interruption of the oesophagus (Nixon and O'Donnell, 1976), is usually associated with a fistula or connection with the trachea. Although this condition was described by Thomas Gibson as early as 1696 (Cywes et al, 1976), only a few attempts at correction were made in the 1800's. Operative procedures were seriously commenced only in the second decade of the 20th century, with the first survivor following staged repair in 1939 and shortly after that in 1941, after a primary procedure.

Oesophageal atresia is a relatively common anomaly with an

incidence most often quoted as 1 in 3000 live births. Although there are anatomical variations, the type most frequently encountered (85-90% of patients), is atresia with a fistula between the distal segment and trachea (Cywes et al 1976), as depicted in fig.1(a).

1. Aetiology

Although a number of theories have been put forward, the aetiology of oesophageal atresia is still unknown. The supposition is that multiple factors in the embryonic stage contribute towards this anomaly. Despite the fact that several familial cases have been reported, no true genetic origin has been demonstrated to date (Cudmore, 1976).

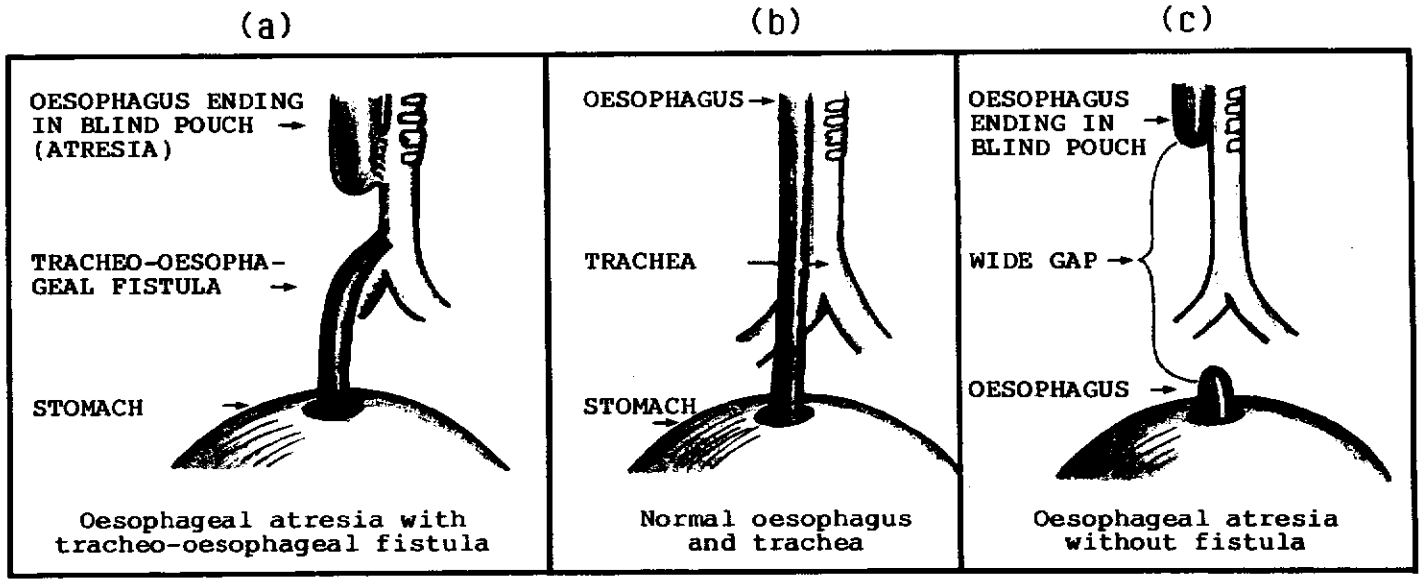
2. Clinical features

The presence of maternal hydramnios (an excess of amniotic fluid) should alert staff to the possibility of oesophageal atresia. This is especially applicable in oesophageal atresia without fistula (fig 1(c)) where the ingestion of amniotic fluid by the foetus is not possible (Cywes, 1976).

Because infants with oesophageal atresia are unable to swallow their saliva, they frequently exhibit a wet, frothing mouth soon after birth. As saliva becomes excessive, the baby may cough,

FIGURE 1

SOME ANATOMICAL VARIATIONS OF OESOPHAGEAL ATRESIA



choke and experience breathing difficulties. These symptoms are aggravated when feeding is attempted. The milk which fills the abnormal oesophageal pouch, soon spills over into the trachea and a real danger of aspiration into the lungs exists. Reflux of stomach contents via the fistula to the lungs is an additional risk. The aspirated gastric juices may cause chemical pneumonia which often only manifests itself after operation, with consequent risk to the infant's life.

On suspicion of oesophageal atresia, a nasogastric tube is passed to the blind sac and accummulated saliva suctioned continuously until the patient is admitted to a treatment centre where radiological confirmation of the diagnosis can be reached.

3. Treatment

The time and type of operation i.e. primary or staged repair, is determined by prematurity in about 30% of cases and the presence of associated anomalies in about 50% of cases (Cywes, 1976).
Full-term, well infants are surgically treated by ligation (tying off) of the fistula and primary end-to-end anastomosis of the oesophageal segments. This means that the two ends of the oesophagus are joined in one single operation. In pre-term infants or those with associated anomalies, a fistula ligation is performed and the patient maintained on a gastrostomy. (The latter is a surgically created opening through the abdominal wall into the stomach for feeding purposes). The definitive corrective surgery can then be done at a later stage when the infant is stronger. Where the distance between the blind ends of the oesophagus is too

great to be joined, (see fig 1(c)) a segment of colon is interposed during the final operation. Possible post-operative complications include a leak at the site of anastomosis requiring surgical repair or a stricture requiring dilatation under general anaesthesia. However, with improvement of preoperative and postoperative intensive care most oesophageal atresias are now repaired primarily even in the pre-term or compromised baby. Various manoeuvres have also been devised (e.g. myotomies) to obtain increased length of the upper pouch to enable a primary anastomosis.

4. Prognosis

Prognosis of oesophageal atresia depends on the factors mentioned above as well as the weight and physical condition of the infant on admission to the hospital. On this basis patients with oesophageal atresia are currently classified into three groups, reflecting progressive degree of risk, as well as influencing the nature of management:

Group A - birthweight more than 2,5 kg and well

Group B_1 - birthweight 1,8 - 2,5 kg and well

B_2 - birthweight greater than 2,5 kg moderate pneumonia and congenital anomaly

Group C_1 - birthweight less than 1,8 kg

C_2 - birthweight greater than 1,8 kg but with severe pneumonia and severe congenital anomaly

According to the above classification, the percentage survival at Red Cross War Memorial Children's Hospital was compared to the series of Waterson et al, which was respectively as follows:

Group A - 95% and 95,8%;

Group B - 68% and 62,5%; and

Group C - 6% and 36,4% (Cywes et al, 1976).

Bearing the different classifications in mind, an overall survival rate of 65% was reported for the 86 patients admitted to Red Cross War Memorial Children's Hospital over the twelve year period 1963 - 1975. Presently the survival rate of full-term infants without pneumonia or other associated anomalies is in the region of 90% or according to some series 100% (Koop et al, 1975).

5. The psychosocial aspects of oesophageal atresia

Illingworth and Lister (1964) were of the earliest authors to suggest a long term effect specific to oesophageal atresia. Included in their case study of 9 children with feeding problems, were 3 patients with surgically corrected oesophageal atresia who had been fed by gastrostomy for prolonged periods pending definitive operations. In all cases the presenting problem had been refusal of solid foodstufs, failure to chew and vomiting. The researchers suggest that these difficulties are related to a "sensitive period", which is the optimal time for a stimulus (i.e. solid foods) to be given, after which time it becomes increasingly difficult to learn the appropriate behaviour. Later, a "critical period" is reached, when a particular pattern of behaviour can no longer be learned. Although the average age for developing chewing ability is given as 6 months, individual maturation and aptitude is stressed. In oesophageal atresia this sensitive period may well have been missed by infants.

In 1977 Dowling studied the development of 7 infants with oesophageal atresia. Two of the studied children who commenced oral feeding during the appropriate period, differed from the remaining 5 who demonstrated the following deficient qualities: forcefulness was lacking in their development, which was already late and slow in advancement. Extreme encouragement was further needed to foster persistence in the child to master a new activity. At the same time little pleasure was shown in new forms of interaction with people or objects. Known activities were also not adequately extended towards new conditions and regressed easily in the face of minor physical illness or other hinderances. Overall their behaviour was described as lacking motivation, vitality or exuberant pleasure with the environment. Although at the age of 4 to 5 months the infants showed preferance for their parents, emotional response was unenthusiastic and tenuous. The authors reasoned that evidence from case histories failed to explain these symptoms in terms of prolonged hospitalization, parental deprivation or neurological lesions. The conclusion was that "the physical act of oral feeding provides an important stimulus and organizing force for a variety of developmental accomplishments" (p.252). Driven by a hunger need, behaviour is extended to other activities: in 5 of the oesophageal atresia children this stimulus had been absent or impaired. This study was however limited to early infancy, which precludes findings on the duration of such symptoms.

In an earlier study by Gibson (1965) projective techniques including Draw-a-Person Test, Wishes and Fears Inventory and the

Childrens' Apperception Test, were utilized to measure emotional disturbance in 9 oesophageal atresia patients with an age range of 5 - 8 years. No significant difference could be found when compared with a control group matched individually for sex, age and intelligence. In view of the long periods of hospitalization and gastrostomy feeding, this finding was somewhat unexpected. The author, however, speculates that mothers with a good relationship with the hospital would be more likely to return for research purposes. At the same time the parents of patients with problems would be less willing to subject them to behaviour evaluation. Bias due to the selective factor in the experimental group could therefore have been instrumental in the achievement of this favourable outcome.

More recently Dera et al (1980), studied groups of children with congenital atresias who had undergone neonatal surgery, mainly for oesophageal, anal and duodenal atresia. Within the study age group 3 - 6 years, those patients where operative procedures were not yet complete in the oesophageal atresia and anal atresia groups, some disturbances in psychosocial development were demonstrated. Separation anxiety and disturbances of contact reached statistical significance on a relationship inventory conducted independently by a paediatrician and psychologist for these diagnoses, whereas the duodenal atresias yielded normal scores. The Stanford-Binet-IQ-scores were within normal range for all groups although at the lower end. This tendency was explained by the fact that the majority of patients had not yet attended kindergarten and were resident in country areas. The most notable result is the absence of aggressive behaviour for all the groups studied.

In summary the authors reasoned that the frequency, duration and early separation between mother and infant, together with the frustrated feeding relationship caused by oesophageal atresia, were major contributing factors to their findings.

By means of a multiple choice questionnaire, Koop et al (1975) investigated the adjustment of 31 families where patients were operated upon between 10 - 25 years previously. Separation varied from 2 weeks to a maximum of one year and four months. Most parents were willing to admit to problems in the past, but seldom in the present. Financial difficulties rarely occurred. None of the mothers had been gainfully employed and tended to feel socially isolated, especially in the patients' first few years of life. This was the time of most acute problems with difficulty in swallowing, choking and the brassy cough, while parents seldom left the patient with a babysitter. The authors recommend that special efforts be made to convey the fullest possible information to mothers immediately after diagnosis has been established. They sound a note of warning against "protecting" the mother and feel that urging the soonest possible visit to the infant is of vital importance. Furthermore long term follow up clinics should be attended by both parents to facilitate sound communication about coping with oesophageal atresia in the home.

The foregoing discussion indicates that formal test results have yielded no clear indications of unilateral psychosocial or intellectual disturbance due to oesophageal atresia. Mediating variables such as mother-infant seperation and transient developmental disturbance caused by disrupted hunger-satiation patterns have been suggested.

B. HIRSCHSPRUNG'S DISEASE (Congenital intestinal aganglionosis)

The Danish paediatrician Hirschsprung, was the first to recognise in 1886 that a congenital malformation might be the underlying cause for a condition which was previously mistaken for acquired megacolon (abnormally large or dilated colon). In the early 1940^{s} the belief was held that the illness was caused by the absence of peristalsis in the aganglionic segment of the bowel. Since 1948 a number of studies showed that the disease was due to an inborn absence of ganglion cells in the involved bowel of the large intestine (Lister and Rickham, 1978).

Ganglion cells are normally present throughout the entire length of the gastrointestinal tract. Functionally these cells move the gastrointestinal contents in an aboral direction (from the mouth to the anus) by transmitting continuous peristaltic waves. In Hirschsprung's Disease the absence of ganglion cells in the affected bowel results in stasis and an accumulation of stool immediately proximal to the aganglionic area. Megacolon therefore develops in the healthy bowel immediately preceding the aganglionic zone which in turn remains narrow and contracted.

1. Incidence and aetiology

The exact incidence of Hirschsprung's Disease is not known. It is seen somewhere between 1 in 2000 (Rickham et al, 1975) and 1 in 10,000 live births (Nixon and O'Donnell, 1976) and is more common in the male than the female by a ratio of 4:1. When occurring in females, greater lengths of bowel are affected. Familial cases have been reported but the genetic origin has remained unclear and

environmental factors appear to play a part in aetiology. A statistically significant association with Down's syndrome has been reported (Lister and Rickham, 1978).

2. Clinical features

The presenting symptoms of Hirschsprung's Disease may most frequently be seen in the first 24 hours or first few days of life, depending in some ways on the length of bowel involved. Infants with longer segments of affected bowel usually present earlier. The most common symptoms are constipation, vomiting, reluctance to feed and a distended abdomen with episodes of diarrhoea. A characteristic symptom of Hirschsprung's Disease soon after birth, is the failure to pass meconium (the first dark green intestinal material passed by a full term infant). In the older child mild malnutrition may be present at diagnosis. Diagnostic procedures include, inter alia, rectal pressure studies, barium enema and rectal biopsies with histology.

3. Treatment

Treatment is aimed at relief of the obstruction and distension which, if left, can lead to mucosal gangrene and death of the infant. In untreated cases the mortality is about 40% in the first year of life (Nixon and O'Donnell, 1976). Bowel wash-outs are given as a temporary measure, followed by colostomy. When the patient is between 6 - 12 months of age a bowel resection and pull through operation is done. Great care is taken not to leave any aganglionic bowel which will cause a relapse, yet at the same time to leave maximum healthy bowel to counteract symptoms like diarrhoea and faecal incontinence. The surgical procedure

therefore involves bringing normal bowel down to the anus.

4. Prognosis

The overall mortality in neonatal Hirschsprung's Disease is 20% or more (Lister and Rickham, 1978) with delay in effecting relief of faeces as a major underlying cause of complications.

In Table 1 the overall results of 80 patients treated at Red Cross War Memorial Children's Hospital during the period 1952 - 1965 is depicted (compiled from Louw and Cywes, 1976). Parents' rating of results will be discussed in Chapter 6.

TABLE 3

RESULTS OF PATIENTS TREATED FOR HIRSCHSPRUNG'S DISEASE

RESULTS	N	%
Early		
Post-operative deaths	4	5
Complications	12	15
Temporary dysfunction	21	26
Late		
Persistent dysfunction	14	18
Secondary operations	6	8
Late deaths	2	3
Good final results	66	83

Post-operative problems include enterocolitis (inflammation of the intestines), leak at the site of anastomosis and some degree of incontinence and faecal impactions. Apart from the trauma of giving birth to a child with this life threatening anomaly, with its subsequent hospitalizations and surgical procedures, the main issue at home becomes the handling of these symptoms.

5. Psychosocial aspects of Hirschsprung's disease

No literature dealing specifically with Hirschsprung's Disease was available in this respect. Some authors (Tejani et al, 1978) include patients with this diagnosis in their sample when discussing the effects of gut surgery. It is hypothesized that psycho-social aspects will be largely similar to the findings of the anorectal malformations discussed later in this chapter.

C. ANORECTAL MALFORMATIONS

Over 30 different forms of anorectal malformations have been described (Cook, 1978) ranging from minor deviations of the anus to complicated, multisystem aberations of the rectum and anus. Early surgical intervention was recorded in the seventh century when Paul of Aegina, a Byzantine physician, thrust a bistoury through the perineum to open the bowel. Increased knowledge of the anatomical basis of the different anorectal malformations resulted in the present sophisticated surgical procedures tailored to the exact pathology of each type of anomaly. Yet these anomalies remain some of the most difficult to treat to satisfaction (Louw et al, 1971).

1. Incidence and aetiology

Anorectal malformations are amongst the most common of the serious congenital anomalies. The generally quoted incidence is 1 in 5000, but varies from 1 in 1800 in the R.S.A. to 1 in 10,000 in France (Cook, 1978). All comprehensive series report a preponderance of males. Although some familial cases have been documented, most appear sporadically.

The aetiology of anorectal malformations is not known except that there is an arrest or deviation in the normal embryological development of the anus and rectum.The wide variety of anorectal malformations are broadly classified by the stage of arrest as "high", "intermediate" or "low". The high anomalies are more frequently associated with urogenital abnormalities, and have a poor prognosis for complete, normal bowel control even where excellent surgical reconstruction of the anus may be achieved.

2. Clinical features and treatment

When, on examination of a newborn, the anus is not present in the normal site, further clinical examination may detect an abnormally sited opening through which meconium escapes. This opening may be in the perineum, vulvar area, vagina or where males are concerned, in the urethra. This will determine the nature and complexity of the surgical procedures required. Some low anomalies viz. in females with a vestibular fistula may be treated with initial dilatation, followed by a back cut operation or a translocation to the normal site. The high anomalies are always serious. Severe

obstruction, the commonly associated abnormalities of the vertebrae and upper urinary tract and often defective pelvic musculature (Louw et al, 1971) are complicating factors. Such cases are initially treated with a colostomy to relieve obstruction. At 6 to 12 months of age reconstructive surgery is performed in an attempt to establish faecal continence.

In a series of 287 patients treated at the Children's Hospital from 1954 through 1969, high anomalies accounted for 40% of cases, low anomalies 40% and intermediate anomalies 15% (Louw et al, 1971).

3. Prognosis

The mortality for low lesions has been quoted at 12% and in high lesions 34% in some series (Cook, 1978). At Red Cross War Memorial Children's Hospital, this figure for high lesions was 25%, in 16% of cases deaths being due to associated anomalies and 6% to delay in diagnosis (Cywes et al, 1971).

Assessment of 66 patients with high lesions three years after corrective surgery, yielded 41,8% good, 34,5% fair and 23,7% poor results, mainly based on degree of faecal continence (Cywes et al, 1971).

Post-operative management requires co-operation between the paediatric surgeon and parents, especially between the second and 5th years of the patient's life. Prior to colostomy closure, anal dilators are introduced to prevent stenosis (Davies et al, 1979).

After colostomy closure, parents are expected to continue with anal dilatations, as well as managing the skills of preventing skin excoriation in the face of faecal incontinence. Equally, faecal retention due to constipation has to be prevented by the parents watching the patient's diet and at the same time administering laxatives in an effective way. In this they are supported by a stomatherapist.

Between the third and fourth years of life, patient training to acquire anal continence, and biofeedback manoeuvres are commenced. Since these children have never experienced the advantages of continence, it may well not have been missed by them (Davies et al, 1979), which means that motivation to master a totally foreign experience has to be generated. Ultimate success of operative procedure,as measured by continence, is therefore in a great measure dependent on parental attitudes and the co-operation of the patient.

4. Psychosocial aspects of congenital anorectal malformations

Congenital anorectal malformations have as yet missed the attention of psycho-social research (Mies, et al 1978). Yet it has long been known that what has been accepted as good results by treating personnel, has often been greeted with less enthusiasm by parents (Woolam, 1964).

The earlier mentioned study by Gibson (1965) included 5 patients with imperforate anus, whose projective test results yielded no statistically significant differences from an individually matched control group. At most, trends were directional, in spite of the

fact that two of the three girls had to cope with embarrassing symptoms of feacal leakage. It should be pointed out however that patients were between 6 - 7 years of age and social acceptance had not yet reached primary importance. On the semantic differential scale the mothers of girls with imperforate anus showed significant tendencies to depreciate the patient and to regard the effected organ as inadequate. The author felt that imperforate anus was an anomaly which was difficult for the mothers to accept. This difficulty was further related to the need for dilatation of the anus by the mother where the painful procedure disrupted the mother/child relationship. When factors like hospitalization, separation and gastrostomy had all been considered, it was concluded that the parent's interpretation of the event and their reaction to the anomaly "must tentatively be seen as etiological factors" (Gibson, 1965, p.236).

Valman (1974) addressed the problem of the possible effects of early malnutrition in patients undergoing resection of the ileum in the neonatal period. A complete series of 8 patients who had more than 45cm of ileum resected were compared with the healthy control group and 13 patients with cystic fibrosis. Evaluation was by means of Draw-a-Man Test and school reports. No significant differences were demonstrated. Furthermore those patients in both experimental groups with the lowest scores had all been hospitalized for the longest periods "for definite social and not medical reasons" (p.426). Tejani et al (1978) on the other hand found some perceptual motor defects on the Bender Test in his patients following neonatal gut resection. Neurological dysfunction as possible cause was not eliminated in this sample.

In their research, which included 20 cases with anal atresia, Dera et al (1980) found some disturbance of contact, as well as separation anxiety, in their sample which had an age range of 3 - 6 years. Intellectual development showed no meaningful impairment but kindergarten and school enrolment was often delayed due to faecal incontinence. Only 8 of the 20 patients were fully continent. In some cases it was felt not to be solely of functional origin but related to the painful anal dilatations which upset mother/child relationships, a factor also mentioned by others (Gibson, 1965). Furthermore, handling of colostomies detracted from the pleasureable aspects of mother/infant interaction which in some cases resulted in rejection. An unexpected finding was the somewhat delayed speech development in anal atresia patients compared to the oesophageal atresia and duodenal atresia groups. The authors reasoned that prolonged hospitalization could not be a related factor, as the oesophageal atresia group had the longest stay of all groups. The effect of anal dilatations was tentatively raised again.

The research literature discussed was mainly limited to relatively young age groups and were often highly selected groups. For example only 11 of 19 patients were available for psychometric testing in the study of Tejani et al (1978). In the German study (Dera et al 1980) only patients of normal birthweight with no associated anomalies or unimpaired central nervous systems were included.

D. THE CHILD WITH A CONGENITAL ANOMALY AND EARLY MOTHER/INFANT SEPARATION

The effect of the handicapped child on the family has seen a wealth of research and published material. However the majority of projects have not adequately differentiated between the nature of diagnoses (Mies et al 1978). Thus the terms "handicapped" and "chronically ill" have been used to group together patients of diverse illnesses and handicaps, both inborn and acquired, which may well explain the often conflicting results of different researchers (Mies et al 1978). For this reason it is unlikely that findings on the handicapped child in general can be indiscriminately used as a baseline for the anomalies of the present study. Such material will be discussed together with the findings of the empirical study, as similarities or differences become apparent.

Certain factors have nevertheless emerged repeatedly as mediating or contaminating variables, irrespective of the anomalies studied. Thus separation of mother and infant due to hospitalization and the associated "failure to bond" is ever present in the literature. This is especially relevant to those congenital anomalies requiring early surgical intervention. The present status of research on the bonding phenomenon therefore needs to be briefly highlighted.

The possible effects of parent-child separation has increasingly held the attention of researchers since the work of Bowlby in the 1950^{s} and later (Bowlby 1969). Emphasis in the 1970^{s} shifted more towards the possible effects of separation in the neonatal period

(Klaus and Kennell, 1970; Barnett et al, 1970; Klaus et al, 1972; Hales et al, 1977). Theories were developed about "critical" and "sensitive" periods which were optimal for developing mother to infant "bonding", with the most extensive work in this field being done by Klaus and Kennell. Other authors have taken this a step further by attempting to determine the limits of the "sensitive period" (Hales et al, 1977). Based on this research, growing importance was attached to the nature of contact between mother and newborn. Lack of or interrupted contact between mother and infant, was seen to result in poor bonding, which in turn detracted from the ability and/or quality of mothering with detrimental effects for the infant.

For a number of years the results of bonding research went largely unchallenged, although some projects had not always supported these findings (viz.Collingwood and Alberman, 1979). Richards (1978, 1979) sounded a note of warning that most research in this field had not provided for any long term follow-up. In the 1980^{S} others questioned the validity of projects based largely on animal studies (Herbert et al, 1982, Chess and Thomas, 1982) and at the same time disputed the existence of scientific proof of a "sensitive period". Three extensive discussions of the literature independently reviewed by Richards (1978), Herbert et al (1982) and Lamb (1982) have all critisized the enduring negative effects ascribed to poor bonding and at the same time seriously questioned the implied irreversability of effects.

In view of the fact that the three authors mentioned above have competently evaluated the contributions over time of a very wide range of research projects including amongst others those of full-term, pre-term and small for date babies, it need not be repeated here. Suffice to say that major imperfections in research design were identified viz. inter alia the criteria by which attachment was concluded to be present or absent (Herbert et al, 1982) and researching mainly socially disadvantaged groups with many other existing problems (Chess and Thomas, 1982). Most critics were in agreement that a positive outcome of the bonding research has been the humanization of obstetric and birthing practices throughout the western world. Lamb (1982) however felt that the exaggerated claims made by this research has done little to advance the credibility of behavioural paediatrics in the scientific community. In addition, those parents who were precluded from early contact with their infants, could be made to feel that an important experience had been missed, which would render them incapable of adequate parenting and nurturing of their off-spring. It was reasoned that this in itself could have a negative outcome for parent-child relationships.

Barbara Korsch's more moderate comment on the work of Klaus and Kennell, very adequately summarizes the present status of bonding research: "Moreover, it seems unlikely that the infant's cognitive development, individual molecular behaviours and interactions between mother and infant, length of breast-feeding, or the risk of child abuse would be the outcome variables that would reflect most sensitively the experiences for mother and infant in the first few hours and days of life.

There may be measurable short- and long-term differences between infant-mother pairs that can be attributed to variations in the experience around and shortly after birth, but so far the evidence is not compelling. Nevertheless, 'the absence of evidence is not the evidence of absence,' and we have a great deal to learn." (Korsch 1983, p.249).

SUMMARY

In this chapter the clinical features of the surgically correctable anomalies studied, i.e. oesophageal atresia, Hirschsprung's disease and anorectal malformations were broadly outlined. This was followed by a discussion of the psychosocial aspects of these anomalies as described in the literature. A grave scarcity of research material was found, indicating a very real need for follow up results in this area.

Because of its pertinence to congenital anomalies requiring early surgical intervention, the findings pertaining to early mother-infant separation, were highlighted.

In Chapter 4 a description of the families under study will be given. The findings relating to the first awareness of the anomaly, and the establishment of a diagnosis, i.e. the pre-diagnostic phase with its concurrent difficulties will then be presented.

CHAPTER 4

THE ONSET OF PRELIMINARY SYMPTOMS AND LEARNING THE DIAGNOSIS

In this chapter the findings dealing with parents' first awareness of preliminary symptoms in their child to the time of an established diagnosis is presented. A brief description of the characteristics of the families under study is followed by findings of the specific difficulties pertaining to each of the anomalies as experienced immediately after the birth of the neonate. The differences in the nature and duration of the pre-diagnostic phase for the various types of diagnoses are highlighted.

A. THE FAMILIES

Eighty-three families with 90 affected children of whom 55 (61,1%) were male and 35 (38,9%) female were studied. The gender proportion was similar to that of the study population (See Chapter 1). Seven families each had two affected babies: they were four patients with cystic fibrosis and 10 patients with Hirschsprung's disease, all of whom were included in the sample. The proportional distribution of families in the different anomaly groups was, to a certain extent influenced by mortality.

1. Mortality

Nine patients representing all four diagnostic categories had died before the study commenced. A further two patients with cystic fibrosis died while the study was in progress, bringing the total mortality to

11(12%). The proportional distribution of deaths by diagnosis is shown in Table 4.

TABLE 4

NUMBER OF DEATHS IN THE SAMPLE BY DIAGNOSIS

Diagnosis	Sample N	Deaths N	Mortality %
Cystic fibrosis	17	7	41,2
Hirschsprung's d	30	1	3,3
Oesoph. atresia	23	2	8,7
Anorectal malf.	20	1	5,0
TOTAL	90	11	12,2

As expected the highest mortality was found in the cystic fibrosis group, where age at the time of death ranged between 6 months and 16 years. In contrast mortality for the other anomalies was limited to the neonatal period, ranging between the second day and first week of life. All four were cases with multiple associated anomalies.

2. Family size

Family size as measured by the number of children born to each family in the sample, is shown in Table 5. This reveals an overall mean of 2,7 children per family with a range of 1-7.

TABLE 5

THE NUMBER OF CHILDREN BORN TO FAMILIES IN THE SAMPLE

No of children	Cystic fibrosis	Hirschsprung's disease	Oesophageal atresia	Anorectal malformat.	TOTAL
1	2	7	2	-	11
2	8	9	7	10	34
3	1	5	8	8	22
4	3	2	2	1	8
5	1	2	1	-	4
6	-	-	3	-	3
7	-	-	-	1	1
TOTAL (families)	15	25	23	20	83
	$\bar{x}$=2,5	$\bar{x}$=2,3	$\bar{x}$=3,1	$\bar{x}$=2,8	$\bar{x}$=2,7

When mortality is taken into account, the mean number of living children per family is 2,5 for the sample as a whole, while this figure drops to 2,1 children per family of cystic fibrosis patients.

3. Age

(a) Patients

The age of patients at the time of first interview or at the time of death, is reflected in Table 6.

TABLE 6

AGE OF PATIENTS AT FIRST INTERVIEW

Age in years	Cystic fibrosis	Oesoph atresia	Hirsch. disease	Anorectal malforma.	TOTAL N	TOTAL %
Less than 1	2	4	3	2	11	12,2
1 - 5	8	8	6	8	30	33,3
6 - 12	3	4	12	6	25	27,8
13 - 19	4	5	7	4	20	22,2
20 +	0	2	2	0	4	4,4
TOTAL	17	23	30	20	90	99,9

For the sample as a whole the majority of patients (54%) had already reached school-going age. The contrary was found for the cystic fibrosis group where 59% of patients were under 6 years of age. This incongruity is explained by the high mortality for this group (see Table 4), which arrested age at an early level. When expected age is calculated by ignoring mortality, the proportion of cystic fibrosis patients under 6 years of age becomes 41%, therefore more closely resembling the rest of the sample.

(b) Mothers

The modal age of mothers in all diagnostic categories fell between 30-39 years (Table 7). The mean age was 36,4 years. There were no mothers in the teen-age group and for 63% of mothers the patient had not been the first child born to them.

TABLE 7

AGE OF MOTHERS AT FIRST INTERVIEW

	Cystic fibrosis	Oesoph. atresia	Hirsch. disease	Anorectal malform.	TOTAL N	TOTAL %
20-29	3	4	5	6	18	21,7
30-39	7	10	13	10	40	48,2
40-49	5	7	4	2	18	21,7
50-59	0	2	2	2	6	7,2
60 +	0	0	1	0	1	1,2
TOTAL (families)	15	23	25	20	83	100,0
	$\bar{X}$=35,1	$\bar{X}$=38,0	$\bar{X}$=37,3	$\bar{X}$=34,4	$\bar{X}$=36,4	

(c) Fathers

Fathers in the sample were older, with an overall mean age of 38,9 years (Table 8). The unknown category includes 5 cases where fathers were absent due to divorce, an unmarried mother and one father who died soon after the patient's birth.

TABLE 8

AGE OF FATHERS AT FIRST INTERVIEW

Age in years	Cystic fibrosis	Oesoph. atresia	Hirsch. disease	Anorectal malform.	TOTAL N	TOTAL %
20-29	2	4	4	4	14	16,9
30-39	3	8	9	9	29	34,9
40-49	6	5	6	3	20	24,1
50-59	3	4	2	1	10	12,0
60 +	-	1	1	1	3	3,6
Unknown	1	1	3	2	7	8,4
TOTAL families	15	23	25	20	83	99,9
	$\bar{X}$=40,4	$\bar{X}$=40,0	$\bar{X}$=39,9	$\bar{X}$=37,0		$\bar{X}$=38,9

4. Social class

For the purposes of the present study social class was determined by occupation of the father, in broad outline following the well-known classification most frequently used in Britain. The nature of this classification and its value for the study of children is well described by Davie, Butler and Goldstein, 1972 (pp.2-6). The range stretches from social class I which comprises those professions requiring the highest qualifications to social class V consisting of unskilled manual labour. Social class by diagnostic category is shown in Table 9. Where a male head of the household was absent, the occupation of the biological father was retained as a measure of social class.

TABLE 9

SOCIAL CLASS OF FAMILIES IN THE SAMPLE

Social Class	Cystic fibrosis	Oesoph atresia	Hirsch. disease	Anorectal malform.	TOTAL N	TOTAL %
I	1	1	2	3	7	8,4
II	4	4	9	6	23	27,7
III	6	10	9	6	31	37,3
IV	1	6	4	5	16	19,3
V	3	2	1	0	6	7,2
TOTAL	15	23	25	20	83	99,9

5. Utilization of social work services

Before embarking on the discussion of specific difficulties pertaining to the anomalies under study, it is necessary to know how many families had been supported by social work services when dealing with problems related to the patient and his illness.

Ten families (Schedule 2) in the sample had been seen by a social worker prior to commencement of the study. In two instances services were rendered by community based welfare organizations only and in 8 cases by social workers attached to a hospital. The nature of intervention had been mainly material assistance and largely limited to lower middle class families.

Social work services were most often rendered to the families of patients with cystic fibrosis (25%), whereas the families of oesophageal atresia patients were proportionately least often helped (4%). Ten percent each of the families of patients with Hirschsprung's disease and anorectal malformations availed themselves of social work services. The first hypothesis, i.e. that a smaller proportion of families of patients with life threatening surgical conditions will have been aided by the

services of social workers than the families of cystic fibrosis patients, is therefore upheld, the proportions being 8% and 25% respectively. The chapters following will examine whether a greater proportion of families in the sample as a whole, had experienced difficulties which could have benefited by social work intervention.

SCHEDULE 2

UTILIZATION OF SOCIAL WORK SERVICES

DIAGNOSIS	RES. No.	SOCIAL CLASS	INTERVENTION
Cystic fibrosis (25%)	012	III	Material
	021	V	Material
	023	III	Supportive
	102	III	Supportive
Oesophageal atresia (4%)	040	IV	Material
Hirschsprung's disease (10%)	002	III	Material
	074	IV	Material
	079	III	Material
Anorectal malformations (10%)	067	II	Supportive
	088	IV	Material

B. THE PRE-DIAGNOSTIC PHASE IN CYSTIC FIBROSIS

A diagnosis was rapidly established for roughly a quarter of the patients suffering from cystic fibrosis. For the rest of this group the onset of problems related to cystic fibrosis not only originated prior to the diagnosis, but contributed largely to the emotional hardships of parents. Closer scrutiny of the pre-diagnostic phase and the factors related to it, is therefore essential to the understanding of parental reaction to this disease.

1. Duration of the pre-diagnostic phase

The beginning of this phase is typically defined as the onset of parents' first suspicion of illness in the child (Falkman 1977, p.19). In the present sample however it was found that many parents were unable to recall such a date accurately because suspicion was usually formed in degrees. For the purposes of the present study the pre-diagnostic phase was therefore defined as the interval between the date of first medical consultation for the babies' preliminary symptoms and the date on which a diagnosis of cystic fibrosis was entered in the hospital records.

TABLE 10

DURATION OF PRE-DIAGNOSTIC PHASE: CYSTIC FIBROSIS

Time period	Meconium Ileus	2nd Affected Child	1st Affected Child	TOTAL
0-1 week	4	1	0	5
2-4 months	0	1	2	3
5-48 months	0	1	8	9
TOTAL	4	3	10	17

The pre-diagnostic phase ranged between a few days and 4 years duration (Table 10). Due to this very wide dispersion, the results for each individual child is presented in Annexure B, Table 1. The five children (29%) who were diagnosed within the first week of life were either suffering from meconium ileus (4) or had been the second affected child in the family(1). For these parents a pre-diagnostic phase in the true sense was really non-existent. In

contrast the remaining 12 sets of parents experienced a growing awareness of preliminary symptoms at varying stages in their child. The diagnostic stages reached for families by the time of discharge from the institution where the patient had been born, is shown in table 11.

TABLE 11

DIAGNOSTIC PHASE AT DISCHARGE FROM MATERNITY HOME: CYSTIC FIBROSIS

Mode of discharge	Patients
Transferred to hospital	5
Discharged with suspicion, undiagnosed	6
Discharged without suspicion, undiagnosed	6
TOTAL	17

At this stage roughly a third each of this group had either been diagnosed, had become suspicious of ill health, or had not in any way been alerted that something was wrong. The difficulties experienced by each of these groups during admission to the institution where the patient was born, will be briefly discussed.

2. Difficulties experienced at the institution of birth

(a) Infants directly transferred to a hospital

Direct transfer to a hospital was effected for 5 patients. All mothers of patients with meconium ileus (4) had a vague suspicion that something was wrong, as the patient did not look healthy. By this time, however, the staff had already arranged for paediatric consultation and parents concerns did not include the establishment of a diagnosis. The one remaining child in this group was directly transferred to a hospital because an older sibling had died of cystic fibrosis. This mother's suspicion had not been aroused prior to diagnosis.

The major problem for these mothers had been the absence of the neonate and the feeling that his life was in danger. All mothers discharged themselves early from the maternity home for these reasons.

(b) Patients discharged undiagnosed, but with suspicion of ill health

Six patients were discharged from the maternity home undiagnosed although staff and all parents had been uneasy about preliminary symptoms. None of the children had reached birthweight on discharge, even though some remained in the maternity home after the mother had been discharged. The histories reflect an initial happiness at giving birth to a normal baby which was gradually clouded by doubt. Yet no constructive action was taken to clarify the position at this stage.

001
"Celia was a very wanted child from the beginning. I already had 3 children and then decided to stop working and have another baby. She was born in the Santa Home, but somehow they did not bring her to me straight away. When all the mothers were given their babies at 6 and 10 o'clock, I was still waiting to see my baby. When the afternoon passed and evening came I was worried to hear that they could not bring her because she was in an incubator. Oh-oh, I thought, heart trouble? You know the way your mind goes. Then I insisted on seeing her and they marched me down to the incubator and there she was, screaming away. Oh well, I thought at least she is alive and kicking. The next morning I did not get her again and then my doctor came and said that he was unhappy because she had a chest infection and her stomach was upset. And later when I took her home, she was constantly wheezy and every month had a chest infection, together with this ravenous hunger which necessitated feeds every two hours."
Celia was diagnosed two years later.

In this group symptoms were handled by staff without informing the parents. Once parents became fully responsible for their infants care, the awareness of symptoms resulted in a medical consultation being sought within days after discharge in all cases. Many consultations and an average of one year elapsed for this group before a diagnosis was established.

(c) Patients discharged without suspicion

Six patients, two of whom were the second affected children, were discharged form the maternity home without suspicion of suffering from cystic fibrosis. In both these instances every effort had been made by doctors at the maternity home to eliminate such a possibility shortly after birth. Both mothers had been unco-operative initially because they had not believed that this would happen to them again. In this way, diagnoses were delayed for four months and two years respectively. The histories clearly indicate an emotional refusal and postponement of the problem, although an intellectual awareness had already developed.

022

"My doctor recommended I have a caesarian section and immediate sterilization. I insisted on a normal delivery because I was healthy and knew that God would not do this to us again - I was going to have a well baby. She was born beautifully healthy and I was beside myself with happiness refusing to be sterilized or to have her tested for Cystic Fibrosis. But within days I knew that she had Cystic Fibrosis. That unforgettable odour of Cystic Fibrosis was present when I changed her nappy. I refused to share this with anyone. Today I will probably react differently, but I was scared and battled on alone. At four months of age she became very ill. My GP walked in and declared, 'this is enough now! Susan will now be tested for Cystic Fibrosis.' Of course the sweat test was positive."

The absence of early suspicion in the rest of mothers while in the maternity home can be ascribed to a variety of factors: Not being fully responsible for the baby, mild symptomatology and mistaking symptoms for possible childhood asthma in an asthmatic family were interacting variables. One unmarried mother showed little interest in the infant, which was neglected and later on removed from her care in terms of the Children's Act. On average more than one and a half year passed for this group before a diagnosis was established.

3. Seeking a diagnosis in the community

Once home, the awareness that their child was not well ensued rapidly for the twelve mothers (70%) whose children were discharged from the maternity home undiagnosed. For the majority a week to ten days passed before medical consultation was sought, with only one instance of a maximum period of nine months recorded. All respondents were in agreement that this had been the most trying phase of the illness, hallmarked by frequent visits to clinics and doctors during the day in an attempt to elicit some assistance with

the child's presenting symptoms. At night children, sometimes critically ill, were handled in relays by parents. Confidence in their adequacy as parents dwindled in the face of an inability to manage persistent symptoms. The histories of five mothers revealed that their competency had in fact been questioned by health personnel.

012

"I was very worried about him because he failed to gain weight and was always ill and fragile. At the clinic they kept on reprimanding me because he had not gained. Then I tried my GP, but he said I had to be patient. At 14 months I did not want to take him anywhere because people in the supermarket were remarking that he was so small. Then I took him to the paediatrician and asked whether he could not be tested for dwarfism. He asked me how tall my husband was and said then that Robert was just born a small chap. Two months later Mrs Martin, a teacher, came to see me. She warned me that people were talking behind my back. They said Robert was undernourished and that I was not looking after him properly. She urged me as a favour to her, to see one more paediatrician. He was the man who had done fat absorption tests on one of her class children. I went, very reluctantly, but he sent us to Red Cross Hospital where Robert was diagnosed."

Similarly when immigrating to the RSA with their fragile 3½ year old daughter, Susan's parents' abilities had already been seriously doubted abroad. A social worker had been requested to visit the family with the view of investigating child abuse, as a possible cause for the infant's failure to thrive.

110

"The hot South African climate left Susan lethargic almost exhausted. Before taking a holiday here I again called in our GP to make sure that she would stand the journey to the north. He urged us to go, stating it would do both Susan and myself the world's good. At the same time I was persuaded to make an appointment to see a psychiatrist on my return for my ill-founded anxiety about my child."

During their holiday the patient became dehydrated for which she was admitted to a hospital where a diagnosis of cystic fibrosis was made by a paediatrician. Mrs. S. saw the latter as almost god-like: "He was the first doctor (and I have gone through many) who helped me."

Seeking a diagnosis for cystic fibrosis in the community had been an experience which negatively coloured parents relationships toward the medical profession. Furthermore, once a parent had been labelled neurotic, avenues for securing advice seemed blocked and different ways had to be tried.

107

> "On leaving that morning, my GP said, 'There is nothing wrong with Donald. In a few month's time he will have grown and will be riding his tricycle in the streets. Then you will be sorry for repeatedly calling me out unnecessarily.' Feeling convinced that Donald was going to die in one of those endless nights of incessant crying, insatiable hunger and frequent stools, I cried when he left. Then I phoned a friend who was secretary to Professor A. She arranged for his admission to hospital to give me time 'to pull myself together'. He was diagnosed within two weeks. Four months later he died. At first I hated every doctor including those in hospital. They walked down the passages talking and laughing, while I knew my child was slowly dying in there."

Shopping around for more opinions after having been accused that her child was not being properly fed, was an allergic baby, had measles and had pneumonia on different occasions, finally forced a mother to seek the aid of a family member who was in the medical profession. As a favour to the latter the child was admitted to a hospital but discharged undiagnosed. Only one option remained: A children's hospital. Here the wards were full, but the sweat test done on an out-patient basis yielded a positive diagnosis of cystic fibrosis.

C. THE DIAGNOSTIC PHASE IN CYSTIC FIBROSIS

1. Immediate response of parents to the diagnosis

The immediate response of parents to the diagnosis of cystic fibrosis was either relief (53%), shock (18%), or disbelief (12%). In a few cases (18%) no noticeable reaction could be isolated.

In view of the very difficult pre-diagnostic phase with the concurrent self-doubt about parental competence, relief was not altogether a foreign reaction to the circumstances. Although the histories show that the initial relief was later tempered as parents became aware of the fact that no cure would be forthcoming, all were in agreement that treatment had either effected remarkable improvement in the patient's condition or had served as a much desired support to themselves.

001

> "When Celia was 2½ years old I was just skin and bones and could not carry on any longer. I then decided to go to Dr.J. and tell him everything and insist on every possible test, which he then did. One day he walked in and said to me,'your child has got cystic fibrosis'. It did not really mean anything to me. Actually I was elated, and I thought to myself now at least we can work towards getting her better. They put her on Cotazyme and she grew almost two inches that month. She then became quieter and more satisfied."

Parents in the sample who reacted with shock seemed the least able to grasp facts about the illness and were less likely to seek information to rectify this.

081

"She was in hospital for a month. Two days before she came out, they wanted to see both of us at the hospital. I remember so well, my oldest daughter was sick at home and he had to go alone. He came home from the hospital and just fell in the door and held onto my shoulders and cried. He was so upset and shocked and I kept on saying 'What happened?' and he said 'It's Tina!' I said, 'Is she dead?' And he said no there is something terrible wrong with her and he wanted to explain to me but said that he could not remember the name."

Later in the same interview and five years after the diagnosis, considerable ignorance with regard to Cystic Fibrosis was still displayed. "I cannot understand the terrible fits of coughing she has. Last Christmas we also had a nasty scare with her. Terrible pneumonia all of a sudden which cleared with antibiotics, though."

Two parent pairs reacted with total disbelief when the diagnosis of cystic fibrosis and it's implications were explained to them. Both followed up by an avid search for more information on cystic fibrosis, if only to prove that a wrong diagnosis had been made in their case.

022

"I did not believe the doctors at all. I was sure they were talking nonsense. I proceeded to read and study cystic fibrosis - everything I could lay my hands on. I believed he would not die - they were making a terrible mistake. Even when I saw him reacting exactly like they had predicted, I still thought they were wrong. But he died when he was three years and eight months old."

071

"When the sweat test was positive, we didn't believe it anyway. My husband has a very religious family and although I am not religious at all, I started believing them that nothing was wrong."

No noticeable reaction to the diagnosis of cystic fibrosis could be gleaned from the interviews and discussions with the parents of three patients: two were from social class V, and had taken the news in their stride, neither requesting further information nor responding to an invitation to answer any further questions they might have.

2. How parents were told.

The greater majority of parents (76%) were satisfied with the way in which they were told that the patient had cystic fibrosis, even though the histories revealed varying approaches which could be open to criticism, such as breaking the news to one parent only. The most important aspect was parents' impression of the attitude of the informant, as illustrated by the following excerpts.

096

> "Mandy was three weeks old when they tested her. Poor Dr.Hall, I can still see his face when I came for the results. He sat at the other side of the desk and it was like he did not know how to tell me. I can imagine how he felt. You know I think it was very hard for him to say something like that to a mother."

A similar uncritical attitude was found in a mother who firmly believed that the diagnosis was erroneous, but that the intentions were good.

022

> "It was an emergency operation directly after his birth. I'll never forget the way the paediatrician came into my room followed by a sister with an injection tray. He was still talking to me when I was injected. I could not really follow everything he was saying before I fell asleep. All I remembered then was that my child would die."

All parents who were critical of the way in which they were told, stressed the lack of information given as being an important factor. The information booklet on cystic fibrosis which was handed to them was not well received.

023

> "I was very upset with the way it was done. First of all they just gave me a pamphlet to read and I did not fully understand what they were trying to say in it. I phoned my cousin who is a doctor and asked him to explain it to me."

012

"After the second sweat test was positive, Dr. Johns told us that Ray had a disease which was mainly concentrated in his lungs but that it would not get any better. He gave scanty information. Something about it being hereditary and urged us not to have any more children. I wanted to tell him then that I might well be pregnant already but I did not have the opportunity. A week later when we visited Ray in the hospital a leaflet on cystic fibrosis was handed to us. As my husband and I drove home, I started reading this in the car. It was terrible. It explained that the children die and it gave all the nasty details. It was a terrible shock and I had the most acute stomach cramps immediately. A visit to the doctor confirmed that I was pregnant. For the next seven months I dreaded having another child with cystic fibrosis."

The lack of information was acutely felt where parents were resident in country areas distant from bigger medical centres. Unbridled anger was levelled at the informant for not supplying the relevant information when the diagnosis was explained to them.

071

"I was reading the "Living and Loving" where they talked about Cystic Fibrosis children having physiotherapy. I realised that we had not been told anything about this - simply that she had a pancreatic disease which also involved the lungs. It was only by pressurising that chauvinist that we finally gathered enough information about cystic fibrosis to treat her correctly at home."

All histories indicated that the fatal outcome of the illness was the most difficult aspect for parents to accept when the diagnosis was conveyed. However only one mother reacted vehemently towards the teller.

026

"Dr.Robbins was very blunt. He said Henry has got cystic fibrosis and at most he can live to the age of ten. That got me, no hope left for him whatsoever! So little more information as to what and why. It was really bad. What happened then was that I became more worried about myself than about Henry. I became a total wreck. I knew I was going off the rails and turned to a psychiatrist who admitted me to a psychiatric hospital. I know that Dr.J. would not have broken the news in such a way, he is such a super person. But to say it's uncurable and he's going to die within ten years, that's

rough!".

No criteria were used to evaluate, objectively, the amount of information given. Where the relationship between informant and parents was good, however, even a perceived lack of information was not interpreted negatively. The following history at the same time illustrates the reaction and coming to terms with information on cystic fibrosis. Verbalizing the fatal nature of the illness remained a problem for all parents in the sample.

001

"He told me it was a thickening of the juices and that it was incurable, but he did not really say anything more. He did not say that one day she would ... well, I didn't actually push him on that aspect. Then I looked at Celia and I thought to myself, I would love her, she would love me, I don't want to panic, I'll just live till that day. He said she could become seven or eight and I trusted him. Then I went to the university library and they gave me two books on Cystic Fibrosis. I sat and I saw the photographs, I read everything. I saw the kiddies died. Then I closed the book - I don't want to know anything more. I want to live for today. I did not want myself to go to pieces and cause more trouble for Celia and my other children. I decided to keep myself so busy that there will be no time to think and I will live for Celia and for today. I had done just that. I have been doing that for years now."

D. THE PRE-DIAGNOSTIC PHASE IN OESOPHAGEAL ATRESIA

1. Duration of the pre-diagnostic phase

Oesophageal atresia was diagnosed the soonest of the four anomalies studied. As defined for the purposes of the present study, the pre-diagnostic phase was really non-existent for the parents of these 23 children. All patients were directly transferred from the institution of birth to a hospital. In contrast to the other anomalies, parents were at the same time in no way instrumental in reaching a diagnosis. In all but one case (with H-type fistula) a diagnosis had been confirmed by a surgeon within 48 hours of birth.

2. Type of Oesophageal Atresia diagnosed

To fully appreciate parental response to the diagnosis, some indication of the severity of the anomaly is called for. Table 12 reflects the type of oesophageal atresia diagnosed in the sample according to Waterson's classification. The consecutive categories roughly indicate the progressive degree of risk to the infant's life (see Chapter 3). Two patients died within two days after birth. Both male infants were of the C.2 classification.

TABLE 12

TYPE OF OESOPHAGEAL ATRESIA IN THE SAMPLE
(Waterson's Classification)

Type	Male	Female	Total
A	2	3	5
B_1	4	4	8
B_2	3	2	5
C_1	1	0	1
C_2	2	1	3
Total	12	10	22

N = 22
1 = Unknown

3. Difficulties experienced in the institution of birth

The events immediately following the birth of their child with oesophageal atresia, were recalled in great detail by all the parents in this group. Although traumatic and seen as a crisis because a normal infant had been expected, the majority of parents (78%) felt that everything possible had been done to best deal with the situation. The early separation from the newborn in an institution "where everyone talks babies" had been especially upsetting for mothers. For this reason all mothers except the four who had given birth by cesaerian section, discharged themselves early. Two of the four mothers remaining in the maternity home were heavily sedated. A third received 4-hourly reports on the infant's condition by the matron of the institution. For 43% of the families emergency transport of the patient to a hospital in a big centre was involved. In six cases the father, sometimes accompanied by a member of the nursing staff, was personally responsible for this.

Five parents experienced problems prior to the infant's transfer to a hospital. Dissatisfaction was most often related to the delay in informing parents about decisions made concerning the infant. For example after his wife had given birth by cesaerian section, a father was allowed to go home without informing him about the child's anomaly or transfer to a hospital.

018

"My husband had cleared off thinking the baby was alright, phoning all over the country to tell everyone. The next morning he did not come and see me, because I was still doped. In the meantime the baby had been transferred to hospital soon after birth. Nobody told him, so the whole thing seemed to be traumatic right through."

Delay of information on three occasions ended in the mother not viewing the infant before transfer to a hospital. In retrospect this was a painful oversight as protracted separation followed due to lengthy hospital admission.

069

"She was born at half-past eleven that night. The next morning I saw an incubator with a drip being wheeled past my room. As she had not been brought to me for a feed yet, it occurred to me that this could be my baby and I started feeling uneasy. I asked the nurse and she said 'Didn't you know your baby was on a drip.' I only saw my child again a few weeks later."

017

"Apparently they tried to pass a tube and then realized that she had Oesophageal Atresia. I had only seen her immediately after birth. When the carrier was taken past my room I tried to catch a glimpse of her, but they didn't want to stop and she was flown to Cape Town."

For the oesophageal atresia group the phase prior to the infants' admission to a hospital was distinguished by the urgency of procedures. Yet accounts of the circumstances were characterized by a manifest absence of reproach directed at staff,notwithstanding

the problems discussed above. This phase can be summarized as a crisis which was generally well handled.

E. THE DIAGNOSTIC PHASE IN OESOPHAGEAL ATRESIA

The urgency of early surgical intervention requires that parents give consent for the operation shortly after birth. Where fathers were not immediately available, this was done by the mother alone. At this time the nature of oesophageal atresia was briefly explained and generally well understood. None of the parents had ever heard of oesophageal atresia and especially in the earlier cases they were told that the infant had little chance of survival. As one parent explained:

> 083
>
> "The dread of an operation on such a small infant was only superseded by the awful prospect of his slowly dying of hunger. I would rather see him die during the operation than to see his slow suffering and death."

The majority of parents had understood that their child had a 50-50 chance of survival and had feared for the infant's life. After hospitalization, news of the infant's progress was mostly conveyed by husbands to their wives. In two instances however marital relationships were such that fathers had been unaware of the infant's condition.

Telling the parents about their child's anomaly had often come from different sources and more information was added by health personnel prior to mothers personally visiting the infants in hospital. Most parents were unable to recall who had conveyed specific aspects of oesophageal atresia to them. The nature of the

operation was generally interpreted by parents as an uncomplicated procedure whereby the blind ends of the oesophagus is joined up. The fear of the infants's death was largely focussed on the smallness of the infant and it's inability "to stand an operation." The expectation was that once the infant survived the operation, the problem would be solved. As can be expected the reactions of the two parent pairs whose infants died was somewhat different. This is discussed fully in a later chapter.

In contrast to cystic fibrosis, the emotional content accompanying the explanation of the diagnosis was mild and almost diffuse. Probes during the interview failed to elicit information with any notable emotional content. Only one case (018 discussed earlier) of dissatisfaction related to telling was recorded.

F. THE PRE-DIAGNOSTIC PHASE IN HIRSCHSPRUNG'S DISEASE

1. Duration of the pre-diagnostic phase

For the thirty patients with Hirschsprung's disease, the onset of symptoms with the concurrent suspicion of either doctors or parents that something was wrong, originated within the first two weeks of life. The duration of the pre-diagnostic phase is depicted in Table 13.

TABLE 13
DURATION OF PRE-DIAGNOSTIC PHASE: HIRSCHSPRUNG'S DISEASE

Time period	Long segment N	Short segment N	Total
0-1 week	4	7	11
8 days - 1 month	3	3	6
31 days - 2 months	1	3	4
61 days - 3 3/4 yrs	0	8	8
Unknown	0	1	1
TOTAL	8	22	30

X = 17 days X = 175 days

As a group the patients with long segment Hirschsprung's disease were diagnosed earlier: i.e. between the first day of life to a maximum of 60 days. Half of the long segment group obtained a diagnosis within four days of birth. At the same time 52% of patients with short segment Hirschsprung's disease had a prediagnostic phase ranging between 49 days and 3 years 9 months. The average time required to reach a diagnosis was respectively seventeen days and 175 days for long and short segment Hirschsprung's disease. As these distributions have a very high variance the exact duration for each individual is delineated in Annexure B, Table 2.

2. Difficulties experienced at the institution of birth

Overall 53% of parents reported upsetting experiences during the pre-diagnostic phase. In thirteen (43%) cases hardships were already present prior to discharge from the institution where the patient was born. As most problems had a close bearing on the diagnostic phase reached by the time of discharge, this information

is summarized in Table 14.

TABLE 14

DIAGNOSTIC PHASE AT DISCHARGE FROM MATERNITY HOME: HIRSCHSPRUNG'S DISEASE

Mode of Discharge	Patients
Transferred to hospital for diagnosis	13
Discharged with suspicion, undiagnosed	7
Discharged without suspicion, undiagnosed	6
Unknown	4
TOTAL	30

(a) Infants directly transferred to a hospital

A direct transfer from maternity home to a hospital was made in 13 cases. An unexpected finding was that 7 of these 13 mothers reported difficulties at this early stage. The most prevalent problem was the yet unconfirmed suspicion of staff members that the infant was not healthy. This was seldom verbalized to mothers.

Their own suspicions were aroused however when one or more feeding times passed without their infants being brought to them from the nursery.

059

"She was born the Sunday evening at 11 o'clock and by Tuesday morning I just knew that something was wrong. They did not bring her for feeds. Each time the other babies came but not mine. With the 12 o'clock feed she did not come again but they said nothing. Then I asked and the nurse said: 'Oh the baby is a bit nauseous.' When visiting time came and I still had not seen her, I had the feeling that definitely something terrible had to be wrong. I said to my husband, ' There is something very much wrong with our child!'".

In four instances attempts by staff to establish a diagnosis prior to giving any information to the parents, resulted in prolonged separation of mother and infant. Fears and phantasies of gross deformities were voiced by two of these mothers.

109

"About 24 hours after birth they usually bring the babies to the mothers to feed, but they did not bring him to me. When the sister passed she told me he had a bit of jaundice and also there was some problem but she did not elaborate. The second day I still had not seen him. I remembered then when I was eight months pregnant I saw on my file they suspected that he would have something missing and I was terribly worried. During visiting hours my mom and I sneaked to the nursery to have a look at Ronald. He was perfect to look at but my mother asked, 'Why has he got a potbelly like a little native boy.'"

In a birth by caesarian section, reticence by maternity staff to permit parents' viewing of the ill infant, caused a delay of first contact between mother and child, of two months' duration.

002

"When I came round from the operation, Anne had already been taken to the nursery and I did not see her. I had a glimpse of her in the incubator when they pushed her to the ambulance to fly her to Cape Town. I only saw her again when she was two months old and she already had the colostomy then."

Where an infant died, this lack of sufficient contact between mother and infant was experienced as an irreplaceable loss.

004

"He only lived for two days.I hardly saw him.They only brought him for two feeds - that's the only time I ever saw my child."

In the remainder of cases where a direct transfer to hospital had been made the roles were somewhat reversed: Mothers experienced trouble convincing the nursing staff that the patient was not well. Although disturbing for parents, this period was fortunately of limited duration. Once consultation with the paediatrician had been established, undelayed admission to hospital was effected.

All mothers of transferred patients (13) whether or not difficulties were raised, mentioned a sense of isolation when left alone in the maternity home without the infant. However, these were not recorded under problems as the parents involved regarded this as an inevitable factor related to the special circumstances for which no one could be held responsible. Fathers were closely involved in transfer procedures of the infant in eight cases and shortly after in taking mothers to the Children's Hospital for the first time.

(b) Patients discharged undiagnosed, but with suspicion of ill health.

While in the maternity home the symptomatology of 7 patients aroused suspicion that an anomaly might be present (see Table 14). Six of the mothers experienced feeding difficulties because the infant was unable to suck and had had no bowel action. The major hindrance to a rapid diagnosis was the failure to elicit constructive attention to such symptoms as the histories of three patients with long segment Hirschsprung's disease reveal.

079

"I battled to breastfeed him and at the same time noticed that he had not had a bowel action. The sister said to me I was just too tense and the baby was reacting to it, I had to pull myself together."

063

"Later on the sisters took me to the nursery where they also had difficulty in feeding her. She was then tube-fed. At the time when I went home, no-one had still managed to feed her."

038

"I knew there was something wrong with him, because one just could not feed him. We had a few fights in there, because the nurses said I was not trying and my doctor was not happy. He came every day and asked them to see a stool, which they said had been thrown away. In this way he was discharged without a diagnosis of Hirschsprung's Disease being made."

Only one mother of this undiagnosed group of patients reported no problems, although the staff were anxious about the infant's condition.

074
"After four days the doctor asked me didn't baby's stomach work, so I said no. Later on when I went home the doctor kept on carrying on about bringing the baby for a check-up."

The subsequent history of this mother in social class V, showed an equal unconcern in the face of numerous health crises of this patient. He was diagnosed at 24 days of age as having long segment Hirschsprung's disease.

After discharge from the maternity home a period ranging from two days to five months elapsed before a diagnosis was confirmed for this group of patients.

(c) Patients discharged undiagnosed without suspicion of ill health

The six patients who were discharged without suspicion of ill health and undiagnosed, all had a short segment of bowel involved. No difficulties were reported before leaving the maternity home. Once discharged the onset of worrying symptoms was rapid. However the patients in this group reached ages ranging between 20 days and 3 years 9 months before Hirschsprung's disease was diagnosed.

3. Seeking a diagnosis in the community

A total number of 13 patients with Hirschsprung's Disease were discharged from the maternity home undiagnosed. The normal channels of securing a diagnosis for a sick child were successfully followed by the parents of five patients: Two patients were taken back to the hospital of birth where a referral to a children's hospital was done as the parents could not afford private consultation. Another two patients with minor visible anomalies were referred to Red Cross Children's Hospital for the same reason. During admission parents' complaints relating to Hirschsprung's disease were followed up and a positive diagnosis made. The fifth

patient was referred to Red Cross Children's Hospital by a general practitioner two days after discharge from the maternity home in an acutely ill condition. A diagnosis of long segment Hirschsprung's disease was made at 12 days of age.

Extreme hardship was experienced by eight families in securing a diagnosis. The histories reveal lasting reproach and aggression directed at consultants who were unable to reach a diagnosis or failed to refer the child to a hospital. A referral was sometimes refused on the grounds that the child had a training problem which needed disciplinary action and not medical examination.

008

> "He had constant diarrhoea, night and day which was followed by constipation, terribly so. He was crying constantly and kept on trying to push. I had been to the doctor a few times but one evening when I phoned again he said, 'Go back to sleep and stop worrying about that child, there is nothing wrong with him.' Later he did refer us to Dr. J. a child specialist who said it was a training problem. That evening I gave him three enemas, but they would not come down again and water started coming out of his mouth. The next morning my mother phoned the doctor and simply requested a referral to Red Cross Children's Hospital. In a few days he had an operation."

Where parents had experienced difficulty in handling symptoms which were not only worrying but painful to the child, especially fathers showed almost uncontrolled anger when this was discussed. These emotions were reflected in various ways in the histories, of which one is offered as illustration.

075

> "He could not stool at all, he had endless problems and I kept taking him to paediatricians but they told me that I over react which is upsetting because you know there is something wrong. I mean a mother can detect that there is something wrong with her child. I know because he used to press and press and I tried to help him, I put hot cloths on his backside to try and help him to

> get it out and eventually there was projectile vomiting. Then I got my GP and he came around. By then the child was running a terrible temperature and he referred us to the paediatrician but he said there was nothing wrong. Father states: I begged him for 3 months just to have an x-ray taken of the stomach to see what was happening there but he refused and said that an x-ray was dangerous to a child.
>
> We were then told to give him suppositories. Eventually he passed something grey and hard. It just shows you the terrible effort it took to push it through that part of the intestine which was not working. So you know how much effort that little boy was putting into pressing that through and then that clown wants to tell me that we are only over-anxious parents. After that when he was in hospital for three days he had over 16 x-rays and then that clown wants to tell me that one x-ray was dangerous. I really feel that the child had to go through so much unnecessary pain and suffering before they made a diagnosis. I can tell you the first day I relaxed was when we went to Red Cross Hospital and Dr.R. came in and he said he was taking biopsies of his bowel. I knew then this man was going to find out what was wrong with my child. It was the only time I felt relaxed."

Where a long period of time elapsed before diagnosis, parents had reached the end of their tether by the time the child was finally admitted to a hospital. Staff who were unaware of this inadvertently added to the existing stress or could not understand parents' reluctance to take the child home where immediate admission was not possible.

> 079
> "I got onto the ward and this real old battle-ax of a sister jumped at me and said, 'How can you bring this child here - just look at this rash - he has got measles.' I thought it was the new cereal I had given him that morning which had caused it, but she said, 'Take him out!' When the doctor came and confirmed measles, I cried. All I wanted to shout was, 'Take my baby, fix him, please just fix him!' I was about ready to hand him over to anyone because I just could not handle it anymore. I was like a miserable zombie. I could not think or do anymore. All I wanted to do was sleep."

In one instance the possibility was seriously considered by a mother whether this very difficult pre-diagnostic phase could not be responsible for the still existing lack of rapport between the patient and herself. Other mothers further described cutting themselves off from the patient and his problems in an attempt at personal survival.

042

"I was very depressed because he did not sleep, he cried throughout the night. With the Hirschsprung's disease he was getting constipated and no-one was picking it up. I can remember getting up at night and putting on earphones, just listening to music because I could not stand the crying anymore."

G. THE DIAGNOSTIC PHASE IN OESOPHAGEAL ATRESIA

The first reaction of parents to the diagnosis of Hirschsprung's disease was mostly a combination of shock and relief. As one mother who had experienced a very difficult pre-diagnostic phase explained:

008

"It sounds cruel, but I was delighted to hear that my child had an illness and that they were going to operate. It's difficult to explain but while they were examining him I prayed, 'Dear God, let them find something which they can cut out!'"

Where parents did express their shock at hearing the diagnosis, it was always qualified with the fact that at the same time there was hope of recovery after operation.

075

"It was quite a big shock when eventually they did discover that it was Hirschsprung's disease. But even then it was better to know what was wrong with your child. He showed us on another child what was going to happen to our child and the bag, how it was working. It was quite a shock."

The reactions of parents to being told that their child was suffering from Hirschsprung's disease showed a similar lack of emotional content manifested by the oesophageal atresia group. This can possibly be attributed firstly to the fact that there was hope of recovery. Secondly, the nature of the operation was interpreted as a relatively simple procedure:

087

"The last part of the bowel to the anus had no nerves to help him push, so everything blocks there. So he said we'll cut that part away and join good bowel to the anus, which he did."

The fact that the parents quoted above complained when a year later the young patient had not fully regained faecal continence, illustrates the simplicity of cure expected. At the time of follow-up, normal continence had long since been reached but was equally regarded as the normal if not delayed course of events.

Some perspective is gained from the comments of those parents who had a second affected child. Apart from the difficulty of accepting the same diagnosis for the second time, there was fore knowledge of the practical implications of what they were being told.

076

"Going through it a second time was a terrible strain. I just could not accept it. At least one thing I knew which gave me hope: There was an operation that could help and the child was not going to die. With the first one they also told us what was going to happen, but it was all just words. I don't think I really understood what it meant. You think, oh he'll react a bit and then he will start stooling."

Although more hopeful at the second child's diagnosis of Hirschsprung's disease, the following history illustrates once more the dearth of emotional response to being told about the illness for the first time.

043 + 044

"I was very young at the time and almost unfeeling when he told me she had Hirschsprung's disease. I think even the doctor was worried about my attitude. But the second time with my son, when they said, 'This is Hirschsprung's Disease we are going to operate tomorrow,' I could not wait. I knew he would live and get better."

Only 3(10%) of parents expressed some dissatisfaction with the way they were told that their child had Hirschsprung's disease. In all cases this was directly related to the lack of information given to them. Where a clearcut diagnosis had not yet been made, a waiting period and being told that the doctor was not available contributed to dissatisfaction. All preferred to be told which tests had been done, whether yielding results or not. An excerpt from the history of a patient who was admitted to a larger general hospital, clearly reveals the parental anxiety when inadequate information was given.

035

> "I really expected more of the hospital and doctors. They phoned and told me that they were going to do a colostomy operation. I did not understand the term and merely asked, 'Will my child live?' When they replied positively I was on cloud nine, thinking that would solve the whole problem. When he came out of theatre I saw this blob of cottonwool on his stomach and wondered what was going on underneath. If only someone had explained. It was a traumatic experience which I will not lightly forget."

Two of the dissatisfied parents were from social class II. The remaining one father was from social class V, where the diagnosis had been given to his wife alone. She was unable to convey the facts correctly, which prompted the action described below.

074

> "They said something or other was blocked and they kept him there for six months. My husband asked me what is that kid doing in hospital and I said I think it's the appendix. Then he took off and said he's going in to speak because they are just cutting him up all the time and we don't know what's wrong. Then we went together and he explained that Allistair's backside had no nerves and they cut it away. And he said if you had one child like that then your next children will be like that."

In no instance was any aggression found which was levelled at the person who told the parents about the diagnosis. The obvious lack of knowledge about Hirschsprung's disease found in the mother (social class II) of a

12-year-old, was not considered due to any default of the informer, even by herself.

063

> "In the end the specialist said he could not find the cause of the trouble and referred me to Red Cross Hospital with a list of test results. There they told me what was wrong - something about a narrowing somewhere, but I was too nervous to understand it and it sounded complicated. I still don't know what was wrong with that child, but they did explain it was something with the colon."

The most prevalent feature for the satisfied parents had been a clear memory of a time when the nature of the operation had been explained to them either on a drawing board or by demonstrating on another patient on the ward. The former method was seen as the most positive and least frightening of the two.

H. THE PRE-DIAGNOSTIC PHASE IN ANORECTAL MALFORMATIONS

1. Duration of the pre-diagnostic phase

Anorectal malformations in the sample were diagnosed sooner than Hirschsprung's disease and the pre-diagnostic phase was also less fraught with problems. In 85% of cases some diagnosis had been reached within the first week of life. The remaining cases were all low anomalies and duration of the pre-diagnostic phase had respectively been of three month's, 18 months' and 5 years' duration (see Annexure B,Table 3).

TABLE 15

DURATION OF PRE-DIAGNOSTIC PHASE: ANORECTAL MALFORMATIONS

Time period	High anomalies	Low anomalies	Total
0-1 week	12	5	17
3 mths - 5 yrs	0	3	3
TOTAL	12	8	20
	$\bar{X}$ = 2 days	$\bar{X}$ = 309,4 days	

2. Difficulties experienced at the institution of birth

As can be expected with early diagnosis, the majority of patients 15,(75%) were directly transferred from the maternity home to a hospital. These included all high anomalies and three of the eight low anomalies.

TABLE 16
DIAGNOSTIC PHASE AT DISCHARGE FROM MATERNITY HOME: ANORECTAL MALFORMATIONS

Mode of discharge	Patients
Transferred to hospital	15
Discharged with suspicion, undiagnosed	2
Discharged without suspicion, undiagnosed	3
TOTAL	20

A smaller proportion of these parents (35%) experienced difficulties during admission to a maternity home than those from the Hirschsprung's disease group (43%).

(a) Infants directly transferred to a hospital

Only five of the mothers of directly transferred patients recorded having experienced difficulties in the maternity home. Although of short duration, the principal problem was still related to diagnostic delay, which was mentioned in three cases. Most disturbing to mothers had been the fact that a visible anomaly had only been discovered later by someone other than the obstetrician in one instance only on insistence that a paediatrician be called. All had a feeling that the infant's chances of optimal recovery had been somewhat jeopardized by this delay.

024

"Almost 48 hours later, the matron noticed that something was not normal. It was such a clear abnormality - the baby had no anus opening. That time lapse radically influenced his chances of rapid recovery."

Viewing of the infant before transfer to a hospital accounted for the difficulties of two more mothers in this group. The intensity of emotions attending this wish is clearly illustrated by the following history given 13 years after the event.

032

"They wanted to take him away and explained that it was probably just a skin over the anus. But I wanted him with me first. It is still printed on my mind the picture of him on the bed with me and they were going to take him away in a carry-cot to the landing strip. I remember the sister then covered up his face and that upset me terribly. I became almost hysterical that she covered up his face as if she had already written him off."

The above at the same time illustrates the disproportionate amount of inexplicable emotion evoked by minor actions of staff, due to the crisis experienced by a parent.

(b) Patients discharged undiagnosed but with suspicion of ill health

Two patients were discharged from the maternity home without a diagnosis being made (see Table 16) although the mothers had sought advice on preliminary symptoms prior to discharge. In both cases regret ensued that they had not insisted more on further investigations.

050

"The first time when I went to change her nappy in the nursery I saw the stools were coming out very high up. I asked the sister why is it like that. She said it's newly born, they'll see later on."

070
"On the third day he was very lethargic, not drinking, not passing stools. Two chaps saw him but said to me don't worry - you are so lucky not to have dirty nappies. Other people said go home and enjoy your baby which I did. So I suppose basically it was our fault for not having done something earlier. But then two chaps saw him and said he was fine, there was nothing wrong with him."

The period of delay for this group was considerably shorter (maximum 7 days) than their Hirschsprung's disease counterparts.

(c) Patients discharged undiagnosed without suspicion of ill-health

None of the mothers of the three patients who were discharged undiagnosed with no suspicion of ill health, recorded any difficulties prior to discharge from the maternity home. All were patients with low anomalies and no negative feelings towards staff about lack of diagnosis was expressed at this early stage. If anything, some self reproach was reflected in the histories.

057

"You see her anus is too far forward. She was checked by Dr.Adams, whom I have tremendous faith in, when she was born but she was so small. I never thought there was anything much physically wrong with her, so I did not have her examined further."

3. Seeking a diagnosis in the community

Five patients all with low anomalies were discharged from the maternity home before the anorectal malformation had been diagnosed. Two of them were home for less than a week before being hospitalized. For the remaining three patients, periods of respectively three months, 18 months and five years passed before the defect was isolated.

Two mothers in this group reacted with acute resentment against the medical profession, feeling that consultations they paid for had not been carried out thoroughly.

084

"He never laid a finger up her anus, so of course never found out what was wrong with her."

There was no correlation between length of the pre-diagnostic phase and anger reactions. For instance the two parents who had waited longest for a diagnosis to be finalised in this group reacted only with relief. Both had previously thought the problem to be of psychological origin and during the pre-diagnostic phase had reacted more towards the patient.

057

"I think I went through every stage with her. I went through being nice and trying to coaxe stools out of her, I went through the stage of clobbering her until I just could not stand it any more. Then I thought good, strict discipline - I went through the whole gamut."

In the second case the family had just gone through a divorce and the illness symptoms had been interpreted by the mother as reactions to emotional trauma. In both instances the mother-patient relationship was felt to have suffered injury due to this phase.

I. THE DIAGNOSTIC PHASE IN ANORECTAL MALFORMATIONS

The diagnostic phase was experienced as a crisis by 13 (65%) parents because they had feared for the infant's life.

092

"It was difficult to accept. When I talk about that time, I still feel a lump in my throat."

062
"I was very shocked and very tense but they kept on reassuring me that the baby was strong enough to stand an operation. Furthermore they felt that Cape Town could do anything for a child."

Whereas some mothers reacted to the trauma of the crisis by trying to forget and sleep, others felt that the situation called for the mobilization of all senses to handle the difficult circumstances.

066

"When it was explained to me I got all miserable and depressed, but they tranquilized me. Being a nervous person I would probably have moped on the subject. Well, I slept most of the time, I was drugged."

032
"Prior to the news of his defect I was not the type of person who could cope with crises. They wanted to give me tranquilizers and all the rest of it, but I refused pointblank. That would be crazy, because what would be the point of being doped if I was going to have to make decisions? I did not want the regret afterwards of not being aware of the facts."

In a further two cases mothers realized in hindsight that they had totally underestimated the implications of the diagnosis and had therefore reacted with relative complacency.

033

"The doctor came back to me and said that his anus was covered. I was not unduly worried because I thought that they would simply cut a little opening and that was it. Then my husband came back and said they were first making a colostomy. We did not realize the serious nature of his problem - that developed in time."

In two cases parents responded with clear relief at the diagnosis. Both were low anomalies and a lengthy and difficult pre-diagnostic phase had been experienced.

057

"It was actually a relief because I got this hang-up of being a lousy mother and incompetent the whole works because of this doctor telling me it was purely my feeding her incorrectly that it was a relief to know that it was not me and there was actually something wrong with her and it was not serious you know, and that they could do something for her and at least we understood the problem, we could actually do something concrete about it."

For the remaining three cases, the reaction of one mother was unknown and two had felt that the diagnosis had not really brought relief to the situation.

084

"So it's just you hope it's going to be better. You don't realize it's going to be worse."

Both the latter cases were low anomalies where according to the mothers the size of the anus improved but not the function.

Eighty-five percent of parents were satisfied with the way in which the diagnosis had been explained to them. No reaction to the "teller" could be found even in those cases where some dissatisfaction was expressed. Dissatisfaction in all cases had been related to the fact that the diagnosis and subsequent treatment had not fully cleared the difficulties of the patient , as in 084 cited earlier.

Satisfaction, on the other hand, was closely linked with parents' feeling that the "teller" was sympathetic and had an appreciation of the difficulties facing them.

027

"He said to us, 'You'll have hassles and it's going to be a tough period. And if things are going to go wrong, this child is going to help it go wrong quicker.' He was a very humane person, this doctor, and he said,'If you have problems,if you have hassles with this little boy and you're worried, phone me and I'll see if I can give you a hand.' He was our pillar of strength because we did start getting irritable with nursing that child twenty-four hours a day."

J. DISCUSSION

In the foregoing presentation the problems experienced by parents during the pre-diagnostic phase were highlighted. Some disparity between the four different anomalies worth emphasizing was found.

The duration of the pre-diagnostic phase revealed marked variance not only between the four groups of anomalies, but also within each separate group. For instance a very short duration was found for all oesophageal atresia patients ($\bar{X}$ = 1 day), high anomaly anorectal malformation patients ($\bar{X}$ = 2 days), and cystic fibrosis patients presenting with meconium ileus ($\bar{X}$ = 2,3 days). By length of duration this is respectively followed by long segment Hirschsprung's disease ($\bar{X}$ = 17,1 days), short segment Hirschsprung's disease ($\bar{X}$ = 174,7 days), and low anortectal malformations ($\bar{X}$ = 309,4 days). Cystic fibrosis patients (excluding meconium ileus) had by far the longest pre-diagnostis phase ($\bar{X}$ = 549,3 days). Because deviations off the mean are wide, reference to the tables in Annexure B is indicated. In summary those conditions requiring immediate surgical intervention, irrespective of anomaly diagnosed, experienced the shortest pre-diagnostic phase.

The nature of difficulties experienced during the pre-diagnostic phase was found to be closely linked with the diagnostic phase reached by the time of discharge from the maternity home. Direct transfers from

maternity home to hospital were self-limiting both in duration and diversity of problems experienced in the pre-diagnostic phase. The proportional distribution of phases reached by the time of discharge from the maternity home for each anomaly is summarized in Table 17.

TABLE 17

PROPORTIONAL DISTRIBUTION OF DIAGNOSTIC PHASES: ALL ANOMALIES

Diagnostic phase	CF	OES	HD	ANO
Transferred to hospital	29,4	95,7	43,3	75,0
Discharged with suspicion, undiagnosed	35,3	-	23,3	10,0
Discharged without suspicion, undiagnosed	35,3	-	20,0	15,0
Unknown	-	4,3	13,3	-
TOTAL	100,0	100,0	99,9	100,0

For the largest proportion of oesophageal atresia and anorectal malformation patients, the pre-diagnostic phase with its related problems was of a limited nature. Hirschprung's disease and cystic fibrosis patients were respectively at progressive risk of longer pre-diagnostic phases and at the same time a wider diversity of problems.

Where a direct transfer was effected, two problems had been present in all anomaly groups. Firstly, a lack of communication was found from staff to parents about a suspected defect in the infant. This gave rise to uninformed speculation or, in a smaller proportion of cases, even phantasies of gross anomalies by the parents, especially the mothers. Secondly, attempts by staff to establish some diagnosis often resulted in transfer of the patient without satisfactory viewing of the infant by

parents. Like other researchers, "We gained the impression that ... staff devoted more energy to making arrangements for the transfer of the child for surgical care than they did to dealing with the shocked, bewilderment of the parents." (Walker, Thomas and Russell, p.465). The incidental finding by Dar, Winter and Tal (p.516) that not viewing gave rise to resentment, was confirmed by the present study. Examples of stealthy attempts at seeing their infant, were cited from the case histories earlier in the chapter.

The far reaching effects of this oversight only become apparent when the length of the first hospital admission is simultaneously taken into account. Table 18 shows the mean duration of first admissions for all anomalies. As dispersions are wide, the range is shown in the second column. Reference is furthermore made to Tables 4, 5, 6 and 7 in Annexure B for individual data.

TABLE 18
MEAN DURATION OF FIRST ADMISSIONS (DAYS)

Anomaly	Duration first admission $\bar{X}$ Days	Range
Cystic fibrosis		
Meconium Ileus	48,5	10-113
Other	5,8	0-11
Oesophageal atresia	114,9	16-685
Hirschsprung's disease		
Long segment	100,9	15-186
Short segment	57,8	4-270
Anorectal malformations		
High anomalies	115,0	13-247
Low anomalies	13,0	1-32

The figures indicate that those diagnoses which had claimed the longest period of first hospitalization (High anorectal malformations, oesophageal atresia, long segment Hirschsprung's disease) were also those where patients were most often directly transferred from maternity home to hospital. The same holds true for patients presenting with meconium ileus when compared to the rest of the cystic fibrosis group. For parents of these patients, inadequate viewing was followed by a lengthy period where mothering was not only postponed but initially limited to visiting hours or such times as treatment regimes permitted. The effects of early mother-infant separation was fully discussed in Chapter 3 (pp.58-60) and will therefore not be repeated here. Furthermore the comments of parents gave sufficient indication of the sizeable dimensions of the emotional hardship caused by this separation.

Three additional factors contributed to difficulties of this early stage. Firstly, being left in an environment with other healthy infants, induced mothers to discharge themselves early. Secondly, fear of the infant's life was present in all directly transferred cases, as parents regarded any operation as a major risk for a newborn. The two last mentioned factors were not only handled with remarkable fortitude by respondents but also not recorded as problems per se as it was reasoned that no-one could be held responsible for them. Thirdly, a small proportion of parents in the Hirschsprung's disease group only gained direct transfer of the infant to a hospital by insisting on paediatric consultation when presenting symptoms became too worrying.

Because suspicion of illness at the maternity home had not been meticulously attended to, major discomfort followed the discharge of 35% of cystic fibrosis, 23% of Hirschprung's disease and 10% of anorectal

malformation patients and their parents (Table 18).The histories reveal that initial symptoms were treated but diagnoses were not pursued nor were adequate referrals made. The least problems were experienced when parents had returned to the maternity home where staff were already acquainted with the infant's symptoms and had responded by making a referral to a children's hospital.

Seeking a diagnosis in the community proved to be not only time-consuming but at the same time a confidence destroying quest. When the patients where no suspicion existed prior to discharge are included, this difficulty was faced by the parents of 70,6% of cystic fibrosis, 43,3% Hirschsprung's disease and 30% of anorectal malformation patients. The proportion of diagnostic delay found for our cystic fibrosis patients is well in keeping with the 73% delay reported by others (Burton, 1975). Falkman also noted that for 35% of her sample between 1 year and 7 years 9 months was required to diagnose this disease (Falkman, 1977). In our sample diagnoses were most successfully forthcoming once referral to a children's hospital had been done. Time was lost due to reluctance of GP's to refer to paediatricians and, in turn, their hesitancy to refer to a hospital if they were unable to reach a diagnosis. Some evidence of lasting hostility towards the medical profession was cited earlier. In other cases parental self-doubt was patent or negative feelings were directed at the patients themselves, where symptoms were regarded as being of psychological origin. A positive relationship between speed of diagnosis and social class was established in cystic fibrosis patients in Burton's sample (Burton, 1975). She surmised that parents in the highest social classes (I and II) would not only communicate more effectively with doctors, but would also have greater access to other sources of information which

would expedite a diagnosis. However our results reveal an exactly opposite tendency; parents from the lowest social class (IV and V) experienced the shortest pre-diagnostic phase for both cystic fibrosis and surgically correctable anomalies (Table 19). This discrepancy is best explained by the fact that the parents in social classes IV and V were most frequently referred to a children's hospital because private consultation could not be afforded. Furthermore when correct diagnoses were not forthcoming parents in social class I and II were more likely to accept symptoms as being of psychological origin.

TABLE 19

MEAN DURATION OF PRE-DIAGNOSTIC PHASE OF CHILDREN BORN IN DIFFERING SOCIO-ECONOMIC GROUPS (MONTHS)

SOCIAL CLASS	I & II	III	IV & V
Burton's sample	8,4	15,0	36,6
Present sample: cystic fibrosis	22,6	13,9	13,2
Present sample: Hirschsprung's disease and anorectal malformations	6,1	16,4	2,4

In summary it can be said that a number of factors influenced the duration of the pre-diagnostic phase. The fact that from the surgically correctable anomalies mainly infants with short segment Hirschsprung's

disease and lower anorectal malformations had been discharged from the maternity home undiagnosed, indicates that severity of presenting symptoms is an important variable in speed of diagnosis. Where preliminary symptoms are not severe enough to effect a diagnosis, other mediating variables account for diagnostic delay.

The immediate response of parents to being told about the anomaly of their child revealed both inter and intra diagnostic difference. The parents of cystic fibrosis children respectively described their relief (53%), shock (18%) and disbelief (12%) at the time the diagnosis was explained to them. In view of the fact that cystic fibrosis had the largest proportion of delayed diagnoses of all the anomalies studied, prevalence of the relief reaction was understandable. To a lesser extent, relief was described in the Hirschsprung's disease and anorectal malformation groups where diagnoses were delayed.

The most frequently recorded response of parents in the rest of the sample, was an acute fear of losing the patient. This fear was most often voiced by parents in the oesophageal atresia group (96%) but also found in the anorectal malformation group (65%) and the Hirschsprung's disease group (57%). In contrast, parents of children with cystic fibrosis found it extremely difficult to verbalize fear of the patient's death.

Parental satisfaction with the manner in which the diagnosis was explained to them, is summarized in Table 20.

TABLE 20

PERCENTAGE OF PARENTS SATISFIED WITH THE MANNER OF EXPLAINING THE DIAGNOSIS

	Cystic fibrosis	Oesophageal atresia	Hirschsprung's disease	Anorectal malformation
Satisfied	76,5	95,7	90,0	85,0
Critical	23,5	4,3	10,0	15,0
TOTAL	100,0	100,0	100,0	100,0

By far the largest proportion of parents in the sample had been satisfied with the way in which the diagnosis was given. The most important positive factor was the attitude of the teller: if the teller was sympathetic, parents had remained uncritical even in those cases where only one parent was interviewed, where too little information was given or where the diagnoses were not fully understood.

A second positive factor was the element of hope offered by the possibility of surgical intervention. The nature of the anomaly was mostly well understood, especially in the oesophageal atresia group. Most satisfied parents could recall that the intended procedure had been explained to them either on a drawing board or demonstrated on another patient. The former method was preferred. Some indications were found however that both the anomaly and the operation with its implications had been underestimated by the parents at this time.

The largest proportion of critical parents were found in the cystic fibrosis group. Factors mentioned were a lack of information, being handed a pamphlet on cystic fibrosis and in one case the bluntness of the teller who offered no hope for the patient. Criticism in the rest of the anomaly groups were levelled at the delay in conveying the diagnosis and the lack of information, but was not accompanied by the vehement emotional response verbalized by the parents of cystic fibrosis patients.

CHAPTER 5

THE CONFRONTATIONAL PHASE: COPING WITH THE ACUTE STAGE OF THE CONGENITAL ANOMALY

In the previous chapter the problems surrounding the establishment of a diagnosis were reviewed. In contrast to the difficulties which were outlined between parents and the medical profession during this pre-diagnostic phase, the present chapter describes an altered relationship as was found to have established itself during hospitalization in the acute phase of the illness. Although this phase was by no means problem-free, the difficulties recorded were markedly diminished both in their extent and the intensity of the attendant emotions. The present chapter further describes the concurrent emotional issue of seeking a cause for the defect. At the same time parents were not only dealing with the anomaly in isolation. Therefore other major life events operative in family life during this early period of the illness are presented in order to gain some perspective on the simultaneous demands made on their crisis-meeting resources. In conclusion some reactions in mothers to first taking the patient home after discharge will be examined.

For the purpose of this study, the acute stage will be defined as the period of first hospitalization and discharge, immediately following diagnosis of the anomaly.

A. THE HOSPITAL EXPERIENCE

During the first post diagnostic hospitalization, many practical and

emotional demands were made on parents. In 44% of cases parents were living geographically distant from the hospital, of which 15 (17%) required air travel between hospital and home. Where neonates were admitted, fathers initially visited alone. In such cases conveying the diagnosis to the mother was delayed, often on insistence of the father. The first visit to the intensive care unit was a totally foreign and mostly frightening experience which reinforced their already present fear of losing the patient - a fear verbalized by 65 (72%) of parents. This first introduction to the hospital was remembered 15 years later as follows:

> 032
>
> "You find that your baby needs surgery - immediately. Unless you are very unusual you won't have known about paediatric surgeons. You would not even have known that there was such a speciality dealing with everything that can go wrong with a newborn baby. Somehow at that time the wheels were set in motion and one was swept on and on in its grinding passage."

This marked the beginning of an association with the institution which would take major responsibility for the physical wellbeing of the sick child in this acute phase - an institution with its own set of rules and own modus operandi. In 84% of cases it had been the first experience of admitting a child to hospital. In the words of one respondent:

> 084
>
> "It was absolutely awful ! You don't know what they are talking about at all."

In the continued association between hospital staff and parents, various factors contributed towards the degree of ease with which this phase was handled. Such factors are both hospital and parent related and must therefore be read against the background of the difficulties experienced in the prediagnostic phase (Chaper 4), the other major life events present and the support systems available to parents in the acute

phase of the illness. Some parents fought the system in order to ensure the best possible care for their child and even left when they were unable to obtain satisfactory results.

> 042
> "I blotted my copy book at the hospital a long time ago. When I complained he would jump down my throat and say the problem had come about because they were shortstaffed and doing the best they can. Yet I am sure I was just a typical mother - you try and get the best for your child. In the end I gave up and paid for the services of private care."

Other parents felt that for the sake of their child they could not afford to make any complaints (065,005,100), but preferred to remain positive at all costs.

In total, 36 (40%) case histories contained no criticism of hospital treatment. Although no direct questions were asked, the remaining 60% revealed a variety of difficulties which were raised spontaneously during the course of interviews. Analysis showed that 86% of parents in the highest social class raised some difficulties while none was found in the histories of social class V families. Table 21 indicates a tendency for the proportion of parents experiencing difficulties to increase with social class.

TABLE 21

PERCENTAGE OF HISTORIES RAISING PROBLEMS RE HOSPITAL TREATMENT

Social Class	I	II	III	IV	V
YES	86	61	61	50	0
NO	14	39	39	50	100
TOTAL	100	100	100	100	100

This finding offers two possibilities: either parents in the lower social classes were less critical of hospital treatment or indeed

experienced this with lesser difficulty. As was shown in Chapter 4, patients of social class IV and V had experienced the shortest pre-diagnostic phase mainly due to early referral to a children's hospital.

The obstacles to effective hospitalization discussed so far were very similar for all anomaly groups. However, parents of cystic fibrosis patients seemed less able to move away from the difficulties experienced in the pre-diagnostic phase. This discrepancy might have been related to the fact that diagnosis and hospitalization had not effected a cure and many symptoms remained largely unchanged.

Five mothers (012,023,026,107,110) from the cystic fibrosis group directly verbalized a negative carry-over from the prediagnostic phase. All had been suspected of neglect, abuse or neuroticism before a diagnosis was made. In its most severe manifestation, hospital treatment was seriously jeopardized, as the social work report indicates.

> 012
> "Since R has been diagnosed, Mrs S has not been able to change her attitude. Although hospital staff are doing their level best to accommodate her in the face of rude behaviour, she perseveres with a very aggressive attitude, on occasion discharging R herself because treatment was 'not satisfactory'".

In contrast, where surgery had dramatically relieved the hardship of the pre-diagnostic phase some examples of positive transference toward surgeons were found.

> 049
> "I knew if I needed help I could phone Dr F. I was so grateful to him, because he was a tremendous help. It's his handywork, you see. If you look at her, it's absolutely fantastic. It was almost as if he took the place of my husband - you know one kind of fell in love with him".

Whether negative attitudes stemming from the difficult prediagnostic phase were to a larger extent covertly present, was not possible to determine.

A further objection which was peculiar to the cystic fibrosis group was the upsetting outcome of wardrounds for the older patient, as the following example illustrates:

> 110
> "I am worried about the fact that they discuss the illness in A's presence. The last time he had been to hospital, he had listened carefully to every word they said. All the terms they used he memorized and couldn't wait to get home to a dictionary to see what they really meant. This really had him worried and in a state".

This difficulty was closely related to the fatal outcome of the illness. As will be substantiated in Chapters 8 and 9 parents were apprehensive of all discussion of cystic fibrosis, lest the patient comprehend the terminal nature of the illness.

1. Hospital doctors and parents

It was evident that a large proportion of parents from all anomaly groups were dealing with feelings of uncertainty about their standing and credibility with the medical profession. Two mothers stated that hospital doctors had labelled them as neurotic. Although this was shrugged off, they confessed to the helpless feeling that once this has happened, one would never be taken seriously again. An underlying caution not to become neurotic or to be seen as such, was found in a further seven histories. These parents related incidents of coping at home with a seriously ill child for fear of being told that the patient did not really require hospital care.

103
"Sometimes she was genuinely very sick, but at hospital I think they used to see an overcaring mother. In fact it was not over caring, it was ... if only they knew the turmoil that was going on. I used to think, now do I take her or don't I take her. You know, I had gone through that dreadful thing about her, is there something seriously wrong or isn't there".

In such cases anger was directed mainly at young doctors. In contrast senior medical staff frequently advised an open ward policy whereby admission could be effected immediately on parent request. The security offered by this policy greatly facilitated handling of worrying symptoms at home with far less anxiety. A further 9 histories clearly indicated a gradual development of parental confidence as a direct result of parent/doctor interaction which was both open and reciprocal.

001

"I went to Dr M, very cheeky, I didn't know then who he was. But I got to like him because I could say to him what I saw wrong. Sometimes he would say to me - 'Don't talk a lot of baloney', but he never made me feel a fool. With the other doctors you've got to shut up. With him we could discuss things."

103
"My attitude certainly did not help them, but Dr L. really pulled me up short. I do have a tremendous amount of respect for him. I know that some of the things he said were harsh, but they were true nevertheless. They gave me a better sense of proportion".

Recognition of their state of crisis was greatly valued by parents as the following excerpts from histories indicate.

002

"I'll never forget, because it was so strange, coming from a professor. You don't expect a scientist to be interested in the emotional side. But when L was so desperately sick my wife and I sat on either side of the incubator and just looked at her. He came in and said he wants us to touch her. We scrubbed and put our hands through the incubator sides and touched her. He said we should talk to her, even if she could not understand. He helped us to become positive again. Later on I thought it was sheer stubborness which made us refuse to give up. That pulled her through."

064

> "There again, I think it was prof Z who was the main source of support - he kept us sane at the time. Honestly, many a time we walked out there and felt absolutely relaxed, having gone in all tensed up".

Similar examples can be cited from 10 histories, referring specifically to the intervention of senior surgical staff as a main source of support when parents were subject to severe mental stress.

The one most frequently recorded complaint during hospitalization was the lack of information about progress, test results and treatment envisaged for the patient. It was clear that a simple explanation by a member of staff could prevent the build-up of unnecessary stress in parents.

> 064
> "I think the worst part of it all was when he was in hospital and you would come up visiting in the evening and there was no one you could talk to. If they would only tell you what was going on. Every time you come it was a different sort of scene. One time he would be in an air tent and the next he was in something else."
>
> 010
> "We felt we wanted to know what was going on - I mean she was your child. We wanted to know everything, whether it was good or bad. I don't know whether other parents were not interested, but to us it felt as if we were being avoided."
>
> 079
> "Everytime I visited him and something new was written on his chart, I wondered what new tests had been done on him and what they had found. I wondered whether he cried, whether the tests were unpleasant and if he could really stand these tests."

Fathers especially resented the fact that procedures or tests were regarded to be beyond their understanding and were therefore not explained to them.

> 073
> "I got the impression that the staff did not regard me as a person of intelligence with whom my child's treatment could be discussed. I can confuse any medical person with engineering jargon which they would never understand, but there are also simple ways of discussing it to anyone's understanding."

As will be shown later in the discussion, the extent of inclusion in planning and in being able to do things for the patient tended to correspond positively with the growth of parent's confidence to fulfill the demands of caring for the patient at home. The more knowledgeable parents became about all aspects of the illness or anomaly, the better equipped they felt to face the day of discharge from hospital. It would therefore seem conceivable that the team should reach consensus on a core of information which could then be conveyed to parents as frequently as possible. Examples of such information might include the free treatment of cystic fibrosis, the value of continuous physiotherapy and the existence of the open door policy. Another aspect of information which needs reinforcement, as will be illustrated by later examples, is the genetic or non-genetic basis of the anomalies. The following history depicts the unnecessary delay of important information to the parents of a cystic fibrosis patient.

> 071
> "They did not tell me why the different medications were prescribed for her - simply said it was good for her. They never told me that hospitalization and medicines were free. Our account had run up well over one thousand five hundred rand when the physiotherapist mentioned that cystics were free of charge. You've really got to suck the information out of people in the hospital."

2. Parents and nursing staff

The hardship allied to poor nursing care was seen as a major problem by 20% of parents. Some aspects of the dilemma were described as follows.

> 079
>
> "The young nurses for instance often forgot to put on some Vaseline when changing his colostomy. It became so sore and raw but I would be too afraid to tell them because they might get angry with me. When I am not there, they still have to look after S. I was afraid they might turn against him, so I said nothing".
>
> 042
> "I found it was better to change the colostomy myself, than to complain. The nursing care was appalling".

Where a lack in nursing care could be alleviated by parental intervention (042 above) parents mostly responded by increasing the frequency and length of their visits to the patient. However, in cases where parents visited and found for instance that a drip had run dry (002,043,080) or an oxygen connecting tube had slipped out (107), they felt themselves incapable of handling the situation. For such care parents were totally dependent on hospital staff. The fact that these errors occurred in their absence, destroyed confidence in the hospital and with it the peace of mind when leaving after a visit to the patient.

002

"When we went up to the hospital to see her, I got the fright of my life. The child's head was out there, it was huge. The drip was dry, it was bone dry and there was no one near her. We could find no one. And then people still say one is worrying for nothing !"

047

"I've lost all my confidence in X hospital through this. I do believe it is because provincial hospitals are understaffed, so I can't blame the hospital, I can't really blame anybody, but I am frightened of leaving him there now".

An open policy for parental visiting served well to allay any unfounded concerns for the well-being of a patient at a specific point in time. A number of examples of visits at an odd hour to set the mind at rest can be cited and much appreciation was expressed for the freedom to visit the ward at any hour. However, a disturbing consequence of the open policy was excessive devotion to 'nursing' the patient at hospital to exclusion of time and care for the rest of the family, often accompanied by guilt feelings from mothers for this neglect.

052

"It helped tremendously that I could go up there anytime. It helped because most of the time I would spend all day with him if possible. But then at night I came home because of the other two. They used to go to my mother-in-law during the day. I used to hate to leave him there at night, though".

Although insufficient nursing care on the one hand and open visiting on the other cannot be singled out as causing this problem, it can at least be seen as additional motivation for such behaviour. Trust in the

nursing staff and in their capable care of the patient enabled parents to go home with fewer reservations. As the following illustrates, appreciation for such support from nursing staff was not lacking in the histories.

042
"I continued to breast feed at the children's hospital. I went parents, hospital and home commitments. This equilibrium was easily disturbed by circumstances at hospital with the nature of the nursing care being one of the most crucial factors in determining the outcome. For instance those histories containing no criticism of hospital treatment did not necessarily describe problem-free admissions. However, they were hallmarked by a positive relationship between parents and nursing staff as the following excerpts show.

052
"It was very difficult when he was in and out of hospital so often. He used to get ear infections or a cold and they had to keep on putting the operation off - that was the worst part. Then they were going to close his colostomy and he got gastro over there and I had to bring him home for a while. Then I took him back again and his stitches festered. In this way his first three birthdays were all in hospital, and every time in the intensive care. Well, I'll tell you one thing, they were terriffic. I mean he had a cake and everything. I think the ward I enjoyed the most was intensive care, they were wonderful. We adored sister E, she is a wonderful woman. I still remember the day I met her - she was carrying two children both with colostomies, each on a hip. I shall never forget that".

083
"We had very pleasant times in the children's hospital. During the 22½ months that he was there, I came down by train 23 times including two Christmasses. I stayed in the nurses home and could see him daily. The staff were wonderful - we had many laughs and tears together".

Of all hospital personnel, the nursing staff are most frequently in contact with both parents and patients, therefore largely determining the nature of the hospital experience for them. Eleven histories indicated that, although various aspects of hospital care were criticised, a member of the nursing staff was constantly referred to as lending order and meaning to a difficult period of hospitalization.

3. Parents and other hospital staff

The above type of reference was to a lesser extent found for other members of staff. The common factor was the restoration of parents confidence in their ability to cope with the demands of the illness.

> 078
> "The one person who really helped me was the stoma-therapist. She was newly appointed to help people with the stomas. She said to me, 'I will give you all the pipes and bags you will need and show you how - I am sure we will cope'. She was the first one to put any confidence in me. After everything they said when I was trying to get him diagnosed, she was the first one to make me feel that I was not pretty useless. I was very grateful to her. And when I came back to her and said this worked and that didn't work she accepted it - she actually had confidence in what I told her".

> 099
> "I know they have all given me up. The only one who still bothers with me is the stoma-therapist".

From the foregoing discussion it is clear that parents seemed caught between getting the best possible hospital care for their child while at the same time maintaining a positive relationship with staff. Once parents had abandoned the concern of being typified over-anxious, neurotic or emotional, the establishment of a healthy working relationship became more viable. It was summarized well in the following history.

> 001
> "I don't care about the doctors thinking me emotional. I have come beyond that stage, thinking I am slightly mental. And I have stopped being timid because I was just going backwards all the time."

The histories cited earlier furthermore confirmed that this point was reached with greater ease where staff attitudes reflected trust in and respect for parent abilities and opinions. In a well established doctor-patient relationship, even harsh criticism was tolerated and accepted by parents (see 103 cited earlier). The somewhat unusual extent of parental vulnerability outlined in this discussion gains better perspective when some of the emotional issues dealt with simultaneously are borne in mind.

B. SEEKING A CAUSE AND REASON FOR THE ANOMALY

During the acute phase following diagnosis of the anomaly, speculations as to possible causes for the anomalies were not uncommon. On one level, it was clear that the correct information as relayed by their doctors had been assimilated intellectually, for example, in the surgically treatable anomalies parents mostly understood the cause as being a "freak". However on the second level, these same parents would also seek causes in a wide range of speculations based on their soul searching and on the events preceding the birth of the patient. The two levels of reasoning were not mutually exclusive.

Discussions as to which side of the family the anomaly could have come from, was not mentioned as frequently as anticipated. Only 18% of the overall sample reported such conjecture (Table 21), which was mostly provoked by grandparents of the patient.

106
"My husband's mother said it could not be his child because he could not have deformed children".

TABLE 22

PERCENTAGE HISTORIES RAISING POSSIBLE FAMILIAL ORIGIN OF ANOMALY

	Cystic fibrosis %	Oesophageal atresia %	Hirschsprung's disease %	Anorectal malform. %	TOTAL %
Yes	12	39	7	15	18
No	88	61	93	85	82
TOTAL	100	100	100	100	100

The low percentage shown in the cystic fibrosis group (Table 21), could partly be accounted for by the fact that parents were well aware that both of them had to be carriers. Burton's sample revealed a similar tendency: between 5%-10% parents reported resentment in this respect (Burton, 1975). The question arose whether rumination on familial origin would be more likely to be present if the baby had been exceptionally special to the family. Ten very important children were identified from the histories on the basis that the parents had described them as such. Schedule 3 summarizes the reasons given for their special importance to the family.

SCHEDULE 3

CHILDREN WHO WERE VERY IMPORTANT TO THEIR FAMILIES

	REASON	N
1.	The only possible male to continue the family name (107,052,014,065,045)	5
2.	The only son after 4 girls (064)	1
3.	Would remain the only child: (a) due to mother's age; (091) - 1) (b) waited 8 years for her (059) - 1) 3 (c) father's 2nd marriage, mother's) first child (002) - 1)	
4.	Only grandchild - sickly grandfather of 86 years living only to see him born (013)	1

N = 10

A tendency was found for the above histories to reveal a greater proportion with controversy on familial origin of the anomaly when compared to the rest of the sample. (The numbers were too small for statistical tests of significance.)

In the sample as a whole, controversy on familial origin was mostly limited to the acute phase of the illness and more specifically to the time shortly after the babies' birth when families were together. Thus where diagnoses were made in new-borns, speculation on familial origin was more likely to be present. The highest proportion depicted in the oesophageal atresia group (Table 32) can be explained in these terms. The situation was described as follows by two mothers of oesophageal atresia patients.

018
"The thing was my mother was with us when the baby was born and taken to hospital and she said, 'Well, there are no abnormalities in our family'. And my husband said well as far as he knows there was nothing in his family and he said he must be paying a very bad

> price ! Something he had done during his life. But otherwise he still seemed to think it was the pill I took. Today I don't know how he feels, but at the time nothing could convince him that it was just one of those things".

> 052
> "He is their only grandchild on that side. It was terrible for them, absolutely disastrous. I know my mother-in-law was in a terrific state when he was born. Every brother and sister who she had, sat up the whole night at her house waiting for his operation to be finished. You know my husband had a little brother with aorta atresia, we went into this but his father had destroyed the birth certificate".

The impression was gained that contention around possible cause for the anomaly had been present to a larger extent during the acute phase for the sample as a whole than was reflected in the present figure. However, the issue had subsided and already lost its importance at the time of study. For example in one case where controversy was denied, the mother of a cystic fibrosis patient nevertheless relayed the following.

> 107
> "I literally forced myself to become pregnant because a son was so important for them to continue the family name. He was born with CF which often made me think that they had made far too much of this whole family blood-line business all the time".

Where very special care was taken to fall pregnant with a son, parents verbalized their reservations as to whether this had been the right thing to do.

> 065
> "It was difficult for my husband's father having 9 granddaughters but no prospect of a male heir to the family farm. We had wanted only 2 children but then decided that we simply had to have a son. We often wondered whether this over-determination was right. Perhaps we were wrong with all this planning and carrying on. Maybe that is why things just did not work out".

In three cases wives were outrightly rejected by their husbands for giving birth to an "abnormal" baby. In two cases the rejection was only transient with both marriages intact at the time of study. One of these fathers (066) reacted by not sending any flowers to the maternity home as he had done in the past, refusing to let the family know of the birth

of his son and blaming his wife that she was responsible. The second father (099) and his family blamed his wife, who was an orphan, that this illness "could only come from your miry past". In the third case (106) the rejection was permanent and he left his wife immediately to raise the child on her own. These examples included parents of patients with an anorectal malformation, Hirschsprung's disease and oesophageal atresia respectively. In six histories clear self-blame was expressed by mothers (075,076,078,079,100,103) for example in the following way.

> 103
> "Feelings weren't always consistent, but occasionally I was riddled with guilt that the basic function of my life apparently I failed in. You know, women are there to bear children and rear them properly".

Occasionally mothers excused themselves, stating that they had stopped smoking and drinking as soon as pregnancy was confirmed (014). In other "causes" mentioned, mothers however mostly implicated themselves as being at least partly responsible for the anomaly, for example during pregnancy worrying too much (039,008), having an electric shock (040), moving furniture (037), being involved in a car accident (100),having x-rays (027,092,049) or anaesthesia (046) and leaning over a pottery wheel for extended periods of time (057). Blame directed away from themselves was rare in mothers. Temporary feelings of anger directed at God during the acute stage of the illness were only recorded in two cases.

> 084
> "I tried to say prayers and things, but I couldn't ... God just wasn't there. How could He do that to a child? How could He make somebody not right and cause him such suffering? That carried on for quite a while and then I suppose your mind gets a bit unhurt. I suppose I was very hurt at the time".
>
> 094
> "I think it's a terrible thing to say, but I fought and wrestled with God in prayer. But later I thought bad things happen to so many people in life, why should I have it any better?"

Self-expressed guilt feelings in fathers were almost non-existent. This can be partly explained by the fact that wives had not always been aware of their husband's feelings and most interviews had been with mothers alone. Only one history contained some reference to feeling partly responsible for the anomaly. Here the husband was 18 years older than his wife and 48 years of age when the patient was born, which he felt could reasonably explain the presence of an anomaly.

C. OTHER MAJOR LIFE EVENTS

The practical and emotional difficulties discussed so far cannot be seen as the only occurrences requiring the attention and efforts of families. In addition, the normal routine tasks of going about living had to be performed. Furthermore 36% (32) families were subject to life events which they interpreted as extreme by way of their demands upon family resources, both material and emotional. The verbalized presence of such major life events for the different anomaly groups is depicted in Table 23.

TABLE 23

THE PRESENCE OF MAJOR LIFE EVENTS IN THE SAMPLE (PERCENTAGE)

Present	Cystic fibrosis	Oesophageal atresia	Hirschsprung's disease	Anorectal malform.	TOTAL
Yes	18	26	37	60	36
No	82	74	63	40	64
TOTAL	100	100	100	100	100

The excessive presence of other life events in the anorectal malformation group is difficult to explain. However, the main purpose of

reflecting the above, serves to indicate that parental difficulties in the acute phase cannot be solely ascribed to the patient's illness.

An extensive literature exists on life events and their relationship to stress. Results have for instance indicated that an excess of life events, notably those regarded as undesirable, may precede the onset of depression (Paykel et al, 1969). Other authors have used both the distress caused by the event as well as the extent of life change produced by the event to measure its impact (Tennant and Andrews, 1976, Vinokur and Selzer, 1975). Because such scales are culture dependant, measurement of the present sample was not undertaken. However, when all three factors (i.e. extent of distress, extent of life change produced and undesirability of the event) are taken into account, it becomes clear that coping with the anomaly was in itself a major life event of some magnitude. The nature of other mostly unrelated life events which were present at the same time, is depicted in Schedule 4. As multiple events were sometimes present in one history, the total exceeds the sample number.

SCHEDULE 4

OTHER MAJOR LIFE EVENTS PRESENT IN THE SAMPLE

EVENT	%
Moved home	13
Lost own business/job or changing job	12
Marital problems/divorce	10
Debt/severe financial difficulties	7
Mother's ill health	6
Death of a grandparent	4
Father's alcoholism	3
Father on military duty	2
Father's ill health	1

In a number of cases it was clear that the unexpected presence of the anomaly and its demands, were directly responsible for excessive undesirable life events, as the following illustrates.

002
"I don't know how we coped the first few months we got here. It was disaster because it was costing us money all the time. We arrived here in fact with an overdraft of two thousand rand. We could not sell the business, we could not sell the house. We just gave up everything - we lost everything. They auctioned the house for x thousand rand which was less than the bond so we did not get anything. The business had x thousand rand worth of stock alone, someone offered half the amount - I had to let go".

066
"We were financially embarrassed when he was born not well and it became very awkward. My husband then held down two jobs to try and rectify this. He was working in the ship store during the day, came home and slept till six. From 6 pm to 2 am he was barman at the hotel". (As cited earlier his wife was rejected and held responsible for the anomaly during this time, resulting in severe marital discord.)

In other cases, the sudden presence of a baby with an anomaly turned the existing problems into a state of crisis, in other words, the birth became the proverbial straw that broke the camel's back.

008
"I think circumstances tipped the scales at the time and everything came to a head. I can't say that I'm sorry. I don't think he would have been a good father to the children. At that time things were so confusing and he was drinking heavily during Charlie's operation. Then I fled to my mother's. I suppose if I didn't, we would still have been together".

Some environmental life events were totally unrelated to the patient, but were felt to be taxing family members so heavily that support was also needed in areas other than the patient's illness.

032
"At the time we had a very bad season on the farm, everything just went wrong. When he was born it was a terrible time for us. For instance the first day when B's health took a positive turn, I phoned my husband and told him that I was allowed to touch B for the first time - it was so special. Two hours later he phoned me back to say that a hail storm had devastated everything - there would be no crop that year. Together with my husband's car accident and the growth that I had to have removed, that time really sounded like a bad novel. We just could not see the light at the end of the tunnel because with all this travelling up and down to hospital we needed a new car for which we did not have the money and so it went on".

From the foregoing examples it is clear that support systems were sorely needed by family members during the acute stage of the illness. Against the background of an accumulation of undesirable life events, the

possibility of depression and other forms of emotional ill-health cannot be excluded. At this stage a much anticipated event was at hand: taking the patient home for the first time after diagnosis and hospital treatment.

D. BRINGING THE PATIENT HOME

The majority of parents (74,4%) described the day of the patient's discharge from hospital as one of the happiest moments since his birth. Part of this much awaited pleasure was often the joy of dressing the baby in the yet unused baby clothes at home (043,044) or simply "holding her, and for the first time feeling that she was mine,"(037). The remainder of parents in the sample were less fortunate. Four babies died in hospital during first admission and a further 19 (21%) mothers expressed a fear of taking their children home.

1. Fear of discharge

As Table 24 indicates, the parents of oesophageal atresia patients expressed fear of discharge most often. At the same time, however, this apprehension was rarely verbalized in the cystic fibrosis group.

TABLE 24
EXPRESSED FEAR OF TAKING THE PATIENT HOME (PERCENTAGE)

Fear expressed	Cystic fibrosis %	Oesophageal atresia %	Hirschsprung's disease %	Anorectal malformation %	TOTAL %
Yes	6	43	13	20	21,1
No	94	48	83	75	74,4
Died	-	9	3	5	4,4
TOTAL	100	100	99,9	100	99,9

The histories throw meaningful light on a number of factors contributing towards positive and negative parental attitudes at this stage - some of which were directly related to the type of anomaly.

(a) Cystic fibrosis

Fear of facing the first discharge of her baby after diagnosis, was described by a mother as follows.

> 107
> "When they told me that I could have him home the next day, I left the hospital in a panic. We already had a mist-tent made which was waiting at home. A humidifier, suction machine and air cylinder was then hired from a private firm and taken home. That night we drove from one restaurant to another till finally every coffee bar was closed. At 2 am we woke a friend for more coffee - I just could not go home. I feared that he would deteriorate and die at home. I didn't want him to die at home. If he died in hospital, everyone would know that everything possible had been done for him."

The fact that the baby died in hospital 4 months later at an age of 9 months confirmed that these fears had not been altogether unfounded. However, for the cystic fibrosis group as a whole, fear of discharge was rare. One contributing factor was that due to delay in diagnosis of the anomaly, parents had already become familiar with the handling of symptoms. Patients were at the same time older by the time of discharge when compared to the rest of the sample. For example the mean age of

cystic fibrosis patients (excluding meconium ileus) was 16-17 months at diagnosis.

(b) Oesophageal atresia

In contrast to the foregoing, in the oesophageal atresia group where fear of discharge was most common, patients had been diagnosed as neonates and hospitalized immediately, in most instances for more lengthy periods than their cystic fibrosis counterparts. The mean duration of first admissions for these groups were 114,8 days and 17,2 days respectively. Therefore no opportunity existed for parents to become familiar with handling the patient and the residual symptoms of oesophageal atresia prior to discharge from hospital.

> 064
> "When he was discharged he had had that long stay in hospital and I was nervous. I think it was because we had never come up against the illness whatsoever. But on the other hand, being so near the hospital was a great consolation - we could just nip up there if anything went wrong."
>
> 069
> "That evening before I had to go home with her, I never ever slept. I sat next to the hospital bed and remembered how the nurses had been doing all types of special things for her. And when I asked questions, professor kept on referring to the fact that I had to bring her up as a normal child."
>
> 015
> "I wasn't really that afraid, but she felt like a little matchbox in that blanket. She was very fragile, but eventually gained weight well. I was terribly anxious that something would go wrong and I watched her like a five pound note every day. As she became older, things became better".

The histories further indicated that the intensity and duration of fear not only varied widely but was more personality related than correlating with the severity of symptoms. In one instance (040) the fear of handling the patient at home was so intense that the mother broke down and was referred for psychiatric treatment. The patient was readmitted to hospital. Other parents merely stated:

039
"I thought they discharged him a little too soon. I would have preferred him to stay longer. Obviously I wanted him home, but we both felt sceptical about handling it".

Confidence in their ability as parents was recurrently found in the histories to culminate in more successful handling of the demands immediately following discharge, as the following illustrates.

052
"Never for a moment did I ever doubt that I could do it, for the simple reason that I wanted him to come home so much. After they told me that the oesophagus had cleared, I knew that all I had to do was to know how to work the bag. I went straightaway to the sister and she showed me and I changed it. She said, 'Look I can see that you will be able to do it'. I brought him home and it never worried me ever."

In a few cases parents insisted on taking the baby home, thinking that they could cope. As one mother who was a qualified nursing sister, explained.

091
"I was actually too hasty, I think. The baby should have stayed there longer, because he still had breathing problems. Of course that Sunday I had maternity blues - and that with a child of 40 days old. I of course rushed back to hospital, but they said, 'Never mind you'll cope later on'. I was more satisfied, but the fact that I didn't have anything with me to help him with the breathing made me very nervous to take him home again."

From certain histories it became clear that home circumstances and social background had certainly been taken into consideration before making the decision of when to discharge a patient. For example in two cases (106,083) with the longest period of first hospitalization, desertion by the father, alcoholism and a generally unstable background had prompted extended hospital stay. On the other hand it seemed that "coping parents" were more likely to experience earlier discharge. As in the excerpt above (091) a mother who was a qualified nursing sister explained the difference in handling symptoms at home as opposed to the hospital. Her feeling was that too much had been expected of her too soon.

018
"When they removed his gastrostomy tube and let him suck on the bottle, he did not have a proper suck and I knew I was going to battle with this child. And when I got him I really had a battle to rear that child. When I hopelessly tried to introduce solids he just pushed it around in his mouth and shoved it out again. You see, another thing, when he was in the incubator and also had the gastrostomy they could let him cry and not worry about him because no one could hear him. But of course we were in a flat and you can't let a child cry endlessly. So there were many times when I was frustrated and just left the child to my husband - he is more even-tempered."

The most uneventful discharges, from the parent's point of view, were distinguished by the simultaneous presence of two factors. Firstly, some confidence in their ability to handle the situation at home, backed up secondly, by a source of consultation if an emergency should arise. When living in close proximity to the hospital, the open door policy mostly served as valued security. In other instances the general practitioner was of vital importance. However this was only true where informed liaison between general practitioner and hospital existed - a fact which was true for all anomaly groups.

(c) Hirschsprung's disease

Fear of taking the patient home after discharge was present in only 13% of Hirschsprung's disease cases. However, the histories confirmed that the overriding eagerness to have the patient home, often rendered mothers ignorant of the practical intricacies of nursing the baby. As the following excerpt reveals, difficulties soon became apparant once the baby was home. The absence of apprehension at this time could therefore be ascribed to ignorance in a number of cases.

035
"I was very eager to get him home because I am very fond of children. Both my husband and I adore babies. We were so excited to have our own baby home, that I did not realize what it would ask of me. I think the most difficult was coping with the colostomy. No one at Y hospital had bothered to show me. In the end I made my own plans and worked out my own techniques of keeping the skin dry and well - but what agony to feel so helpless".

Anxiety of mothers, preceding or immediately following discharge of the patient, was most often directly related to handling of the colostomy. This was especially relevant in earlier cases prior to the appointment of stomatherapists or even later cases discharged from hospitals where no instruction had been given.

> 042
> "I was terribly anxious to take him home. Too little guidance was given when he was discharged with the colostomy. I was tensed up because I did not know how to handle this".

In one case (063) a number of discharges had been well handled by the mother. Then followed discharge for the first time with a colostomy after which she fell into a severe depression. She related this directly to the colostomy which had been totally foreign to her and which she was afraid to handle. Careful instruction and feeling competent with the colostomy on the other hand were most often mentioned as positive factors in uneventful discharges. It seemed as if parents would be more comfortable with a list of complicated instructions rather than being left to follow their own intuition in raising a child. As stated in the discussion of oesophageal atresia cases earlier, some informed support available in their own community was of great importance as is well summarized in the following history.

> 003
> "I never asked too much to take her home because I don't know much about nursing and medicines - I can't take that kind of responsibility. But when they said I could take her home, fine, I took her home and they gave me a list of instructions and off I went. I knew I had Dr H to back me up at home. He was one of those doctors who never gave the impression that you were wasting his time".

Armed with some direct instructions, the physical well-being seemed to be regarded as a challenge for some parents (see 109). For others discharge implied competing with the hospital to keep the patient well, which was seen as a threat and therefore feared (see 002).

109
"Put it this way: They showed me what I should do and how to handle him and I was determined to make a success of it all. When I brought him back for the pull through operation, he was really quite fat and they could not believe that he looked like this. It really took some doing to get him like that. My husband made a cage contraption to keep the blanket away from the colostomy. I tried gum, I tried all sorts of things to keep the motion from running onto the skin. I managed to get some stretchy, round bandage that would not hurt him and we kept his hands tied up to keep them out of the way. He had to lie around naked most of the time to keep the skin dry".

002
"We were terribly worried when we first took her home. After months of strict supervision in the hospital, the responsibility to keep her well and healthy was too much. So, to start off with, we kept total records like they do in hospital of bowel movements, food intake - for ages. It was rather a nerve racking period".

As in oesophageal atresia, the length of hospitalization seemed to be related to parental anxiety at discharge. The extended duration of admission was always mentioned by the parents, yet never clearly spelt out why it had raised the level of anxiety.

038
"He was already 8 months old when I got him home for the first time. The first 3 months home was a nightmare. He had to be fed every 2 or 3 hours, 24 hours a day".

(d) Anorectal malformations

A slightly higher proportion of parents in the anorectal malformation group verbalized fear of taking the patient home when compared to the Hirschsprung's disease group - percentages being 20% and 13% respectively. The relevant factors were similar to those discussed for Hirschsprung's disease with fear often being linked to unpreparedness to handle a colostomy at home. Similarly, emulating hospital treatment at home was sometimes found to bring a form of security even where no fear had been verbalized.

049
"I was dying to get her back again and terribly strict about her treatment regime. I kept up the input-output charts so that I knew exactly what was happening".

Other parents only remembered their anxiety and really had no plan worked out but to face each day as it came. A mother, without any support from her husband, explained:

> 066
> "I was petrified - out of my mind and could not tell a soul. But I thought, well, I will take it as it comes. I don't know how it ever worked, because every day was the same - unpleasant".

From the discussion it becomes clear that whether or not fear of discharge was expressed, taking the patient home for the first time implied shouldering a major responsibility for most parents. While some regarded home-coming as a challenge, others saw an uncomfortable threat in the event and still a third group only comprehended the implications some time after discharge. Clearly the majority of parents were looking around for tangible guidelines to follow at home.

The foregoing discussion immediately raises the question of whether the presence of the congenital anomaly and its related aspects in any other way hindered early assimilation of the patient into family life. Some feelings regarded as largely extraneous to bringing a healthy child home, were identified. It was more commonly found in the parents of patients where the diagnosis was made in the neonatal period or during early infancy. As will become clear from the discussion, immediate post discharge assimilation at home was more uneventful for those patients who were beyond infancy.

2. Mother-child estrangement

Mother-child estrangement varying both in intensity and duration was found to be present in the sample. Apart from the casually mentioned feeling of reservedness towards the patient, a further handful of examples were observed where mothers outrightly confessed to the feeling

that the child was not theirs (107,018,047,091,038,044,075,082,027,062) of which three are offered as illustration.

075
"I could not take people asking me how C was, more so because I had to accept my child again when he came out of hospital. He did not feel like he was my baby. I felt more like he was the hospital's baby and I was looking after him for them. He did not really belong to me".

062
"There was one week's break between the time when I had my baby and when I could first come down to hospital. When I saw him after that, I did not feel that I loved him. It was funny but I did not feel that he was mine. It was as if he belonged to the hospital. I felt when I picked him up that I could not say to him 'cootchy-cootchy coo !' I just could not relate to a baby that wasn't mine".

047
"I wanted to give the child love like a mother normally does, once he was home. But then, when I was able to bring him home, I am afraid something switched in my mind again and I turned off. I felt terrible afterwards. I would attend to his need when he cried, but that was all, I could not hold him, I could not love him. I had to learn to love him and it was a good 4-5 months before I felt something for him. I just felt as if he was ... allright he is here, he needs me for certain things - to be fed, bathed, cleaned, etc. and that was all. There was no feeling of great love towards the child. The child was there and I had to look after him and that was all. Slowly, of course, things turned. I think he is a favourite of mine now".

The disrupted mother-child relationship described in the three excerpts above was clearly of a temporary nature and limited to the acute stage. However a smaller proportion of the histories further bore evidence of long term impairment which mothers acknowledged as originating in this acute phase and remaining unresolved through the so-called long term adaptational phase.

In five cases (016,037,064,106,083) histories displayed the other side of the coin. Although no feelings of strangeness were professed by the mothers, they had however, observed these feelings in their recently discharged infant, and had reacted by donning white uniforms in an effort to somewhat replicate the familiar hospital environment with its nurses. All were convinced that the infants were pining for the nurses whom they regarded as their mothers. In one instance children of the neighbourhood were rounded up to further simulate the hospital environment - complete with nurse and other patients. Although very taxing for mothers, the emotional distance of infants in all cases was felt to be short lived, resolving itself within a few days to three weeks time.

3. Death wishes

The often tacit desire that the patient should rather die, was found in some parents who were afraid to take the patient home, in others who insisted that the patient come home "if only to die here", and in those who did not want the patient to die at home. Although no direct question was asked, this information was nevertheless offered by those parents who felt secure enough to verbalize their feelings. In most instances the serious nature of the illness was stressed at the same time to motivate this wish.

> 038
> "I am ashamed to admit this, but there were times when I almost hoped he'd go in the very beginning. We did discuss this, my husband and I. We said, look, if this child is going to be so ill and we might have problems later, rather let him go now and start again. That is a terrible thing to say, but he was not really my child yet. I had never had any problems falling pregnant or anything like that. It would have been easy to have ... I had easy births. I would quite happily have another one rather. That was initially when he was desperately ill and of course I wasn't well. I had a breast abscess and could not breast-feed him".

048
"I was sick myself and I was not allowed to pick her up or anything like that either. So as it was I really was hoping that this baby would not make it".

It was not possible to ascertain the exact proportion of parents who had experienced death wishes for the patient, as the ability to express such controversial and ambivalent feelings differed extensively. Furthermore, it was clear that while the hospital and medical profession had invested so heavily to ensure patient survival, parents found these feelings unpermissable to themselves. More acceptable expressions like having become reconciled to the possibility of losing the patient and projecting an acquiescent attitude in this respect, was a common finding.

059
"I said to my husband, if God wants to take Annie away, then it has to be so. I came to the conclusion that in a case like this, His will had to happen".

In cases where multiple anomalies were present, the options of active intervention or conservative treatment till the imminent death of the patient were discussed with parents. These examples yielded very different responses to the death of the patient which will be discussed in Chapter 9.

4. Wishes to have aborted

Seven mothers (022,018,048,038,075,076,027) lamented the fact that they could not have or had not been helped to abort before the patient was born. Included were two mothers of children with oesophageal atresia, one with cystic fibrosis, three with Hirschsprung's disease and one with an anorectal malformation. The most vehement reaction was found in the mother of a child severely affected with cystic fibrosis where the risk had been known to them.

022
"My cycle was upset and I fell pregnant. But I did not want another child. I just did not want the child. By that time I realized that I could not take a chance in case I had another cystic fibrosis child. Then what drama! The GP said, 'let's remove it', but the gynae said 'You are insane, this is not done. There is a very good chance that this can be a healthy child'. It was not easy. We talked night after night, we cried, we prayed, absolutely torn between two decisions".

In the non-genetic anomalies, no foreknowledge of risk factors existed. Nevertheless mothers felt that bleeding spells should have been heeded as a warning sign that all was not well. Therefore abortion should have been facilitated rather than hindered. However ambivalence was present and some insights were only gained in hindsight, following the difficulties of handling patients at home.

027
"My belief was that the baby was trying to abort until I went to a gynae who said, 'We have got to stop this bleeding, I will put you on to hormones'. At twenty weeks I had stopped and was in bed with my feet up. But obviously it was a means of nature saying this is not right to continue. I think it must be allowed to take its natural course. It's just that earlier I was right behind the doctor - you know the conflicts all the time. If I had known what I had to face afterwards, I would recommend anybody not to stop aborting, truly. If my husband should hear me now ... he adores the child. I still think one should not play around with nature like that if it wants to abort".

018
"I still feel that if I had had a more severe bleed and had the miscarriage, it would have been fine as far as I was concerned. To me, I mean that is nature's way of getting rid of a lot of abnormalities".

048
"My feelings were ... I think it was resentment at not being able to abort because I feel that would have been a far better situation for everybody concerned".

The interplay of the feelings discussed above and the handling of residual symptoms described earlier, became important factors in slowing down the integration of patients into family life. Because the continuance of symptoms varied widely, in some instances extending well into adult life, a more detailed discussion will be given in Chapter 6 where aspects of the long term adaptational phase are highlighted.

E. DISCUSSION

1. During first hospitalization of a patient following diagnosis of a congenital anomaly, it can be expected that a large proportion of parents will be dealing with the fear of losing their child as found in 72% of the present sample. Many were subject to travelling extreme distances to hospital (44%) and for most it was the first experience of hospitalising a child (84%).

2. Parents in the highest social class were most often found to be critical of hospital treatment (86%), a feature displayed somewhat less for the sample as a whole (60%). The most common complaint throughout was the lack of information about treatment, test results and other procedures.

3. The parents of children with cystic fibrosis were most likely to show overt signs of a negative carry-over of problems experienced in the pre-diagnostic phase (29%). Overall parents in the sample had been uncertain about their credibility with the medical profession. However this changed positively where an open doctor/patient relationship was established. The former, together with parental inclusion in treatment of the patient, tended to build up the confidence which was so badly lacking in parents. The excerpts from histories adequately illustrated the profound influence exercised on parents where medical or surgical staff had been sensitive to their emotional needs.

4. Complaints about nursing care (20%) were coupled with a disturbed equilibrium between parent's home and hospital commitments: in such cases too much time was spent at hospital. Positive relationships with nursing staff on the other hand not only facilitated mastery of several difficulties during hospitalization, but also resulted in not complaining even where serious problems had been patently present.

5. Contact with the stoma-therapist gleaned only positive comments from parents where patients had been discharged with colostomies. Two factors in this contact unmistakably eased discharge: restoration of parental confidence in general and more specifically the knowhow in dealing with colostomies.

6. Although being intellectually aware of the correct information about cause of the anomaly, wide speculation on an emotional level was nevertheless found in the sample. The presence of such speculation in itself need not necessarily be a reason for concern. However, the nature of mothers' reasoning tended to internalize the cause of the anomaly, in other words, somehow a family member could be held responsible for its presence. Seen in terms of Hill's conceptual framework (see Chapter 3), the source of trouble or stressor then became intrafamilial and therefore more likely to lead to family disorganization and breakdown. In this regard the views of fathers would be of importance, but were only evident in the few cases where unconcealed rejection of their wives was recorded. The fact that a large proportion of families (36%) were also dealing with other major life events at this stage, accentuates the importance of relieving their crisis meeting

resources of unnecessary stress originating from erroneous beliefs about causes of the anomalies.

7. Direct blame between parents of cystic fibrosis patients was rare in both the present and other samples (Burton, 1975). This was also true of the surgically correctable anomalies in our sample. However, the tendency for the latter mothers to indirectly implicate themselves for causing the anomaly, was present and they could only benefit from repeated reinforcement of the chance nature of the anomalies.

8. Some parents (21%) anticipated the day of discharge with fear and still more started doubting shortly after the discharge the wisdom of having the patient home so soon. The specific practical difficulties pertaining to each anomaly group were discussed. A common trait was the feeling of unpreparedness to handle the situation: parents were looking around for rules to follow at home. Positive factors identified were firstly, confidence acquired during hospitalization, mainly due to staff attitudes and knowledge gained about the anomaly; secondly, the open ward policy where emergency admission could be effected by parental request and thirdly, the support of a general practitioner in the community.

9. On the emotional level apprehension of discharge was coupled with other factors inter alia fear that the patient would die at home. Together with the reciprocal estrangement between mother and child and death wishes for the patient, the foregoing clearly indicates that many parents need both practical and emotional preparation for coping effectively with the first discharge of the patient.

Summarized in broad trends, this chapter clearly reveals that the acute phase of the anomaly, together with the practical difficulties of the patients' first hospital admission and discharge, cannot be seen in isolation. When the effects of, and the reactions to this stage of the illness are considered, the simultaneous presence of other major life events has to be borne in mind. Furthermore it was shown that the abilities of some parents to successfully meet these demands were still being hampered by unresolved issues emanating from the prediagnostic phase. At this time new issues were already at stake: possible causes for the anomaly were being contemplated on both intellectual and emotional levels by parents who were simultaneously relied upon to establish meaningful working relationships with hospital staff. The advent of discharge with the concurrent intricacies of nursing the patient at home, was faced with different degrees of ambivalence, ranging from eagerness to fear. Some evidence of emotional factors militating against easy assimilation of the patient, for example wishes to have aborted, death wishes for the patient and mother-child estrangement, were found.

CHAPTER 6

THE LONG TERM ADAPTATIONAL PHASE: LEARNING TO LIVE WITH THE ANOMALY IN THE HOME AND COMMUNITY

In the previous chapter the factors relating to the first hospitalization of the patient following diagnosis were discussed and the emotional and practical difficulties of first discharge were highlighted. The present chapter deals with the time following such discharge when parents are required to cope, at home, with the patient and his residual symptoms and treatment. This was essentially a time when the major responsibility for the patient's well-being shifted from hospital personnel to the parents at home. This was also a time when parents were seeking avenues of support to fill the void created by the loss of the expert care which the hospital had provided for the patient. These supports had to be found within the ranks of the nuclear family as well as the community.

The study material indicated firstly that the availability of support not only varied widely, but secondly was largely influenced by the attitudes of parents towards the community. Furthermore the needs of the parents on the one hand and their ability to successfully utilise the support on the other, were not fixed but developed and changed over time.

The contents of this Chapter are based on the views of the parents on the current symptoms and the way in which they had been handled up to the time of study. The findings on community reactions to the anomaly and the parent's attitude are presented, and add further perspective to the demands posed by the illness in the long-term adaptational phase.

In conclusion the extent of contact between parents of patients with similar conditions is briefly looked at. The support found within the boundaries of the family will be elucidated in Chaper 7.

A. HANDLING SYMPTOMS AT HOME

Parental reaction to the prospect of raising a child with a congenital anomaly varied between hopelessness at the overwhelming magnitude and seemingly endless duration thereof, to a firm sense of commitment to tackle and successfully overcome the problems ahead. Many parents vaccilated between these two extremes, the determining factor frequently being the current state of the patient's residual symptoms. The initial despondency was verbalized as follows by the mothers of two patients with anorectal malformations.

> 094
> "It was almost too much for me - to think that I had to raise this child. At that moment I felt so totally drained of strength. Would I really be able to handle this? I don't believe I can!"

> 103
> "I was convinced that I was going to have a problem for the rest of my life. I saw no way out of this. I can see myself hating her and still not coping and being stuck with her for the rest of my days - because she was mine."

The nature and duration of the difficulties experienced varied considerably between the different anomaly groups. As will become clear from the discussion, long-term adjustment in families of patients with oesophageal atresia followed a more or less chronological course. However, families of patients with cystic fibrosis showed wide individual variance in long-term adjustment due to the fact that this anomaly varied so widely in severity and time in running its full course. Thus all stages, including long-term adjustment, might well have been experienced, for example in periods ranging between 9 months

or as long as 18 years. These differences complicated the design of a simple base of comparison between the different anomaly groups and should be taken into consideration when reading results.

1. Cystic fibrosis

One of the most difficult aspects of reaching a long-term adjustment to cystic fibrosis, had been the fatal nature of the illness. Whereas most parents evaded talking about the patient's eventual death, there were the few who could verbalize the negative effect of such thoughts on settling down to treating the patient at home. This was described as follows by parents, in the first case a foster mother.

> 021
> "In the beginning I thought to myself, O Lord, what is the use of carrying on? I mean he is going to die anyway! But it helped when I accepted that he's got cystic fibrosis and nothing I do is going to cure it, but everything I do is going to help him."
>
> 107
> "How can one accept a challenge for which you know in advance that you are going to loose? In the end I settled for a lesser goal - trying to keep him well beyond the year that he was given to live".

Perhaps due to the fact that end results could not be successful, many parents tended to devote themselves to controlling the symptoms very meticulously. One mother (001) described "making a career" of keeping the patient well, which at the same time "would keep me so busy that there would be no time to think". Mothers, especially those in the higher social classes, spent a lot of time "researching the illness", experimenting with diets and writing to cystic fibrosis associations abroad for more information in order to better equip themselves to curb symptoms. A few ended up feeling (022) "more knowledgeable about cystic fibrosis than the professionals" (001,022,110,071).

Shopping around for a medical cure of cystic fibrosis after the diagnosis had been made was not found in the sample, although one patient (022) had accompanied the parents on a holiday to Europe where a doctor was consulted as part of routine treatment. Only one father of a patient (012) turned fully to religion and divine healing for a miracle cure but this was counter-acted by the mother who persevered with treatment of the patient.

Although patients in the cystic fibrosis group were discharged with the most extensive and time consuming home treatment regimes, the expected hindering effects on patient integration into family life was again seldom verbalized in the interviews. Such information was found however in those histories taken after patients had died or where a patient had been removed from parental care in terms of the Children's Act. For example a mother referred to the severe effort of keeping extensive records of food intake, urine output and even weighing of stools for a full year following patient discharge home (071). Another parent explained how a taxing 24 hour time schedule had to be adhered to in order to treat the patient, clean mist-tents or suction machines and do physiotherapy three times a day. At the same time an intercom system to monitor patient breathing difficulties at night was set up (107). Both histories revealed that handling of pancreatic and respiratory symptoms had claimed full and almost exclusive attention for respectively 12 months and 9 months following the patient's discharge. In a third example (021) the mother soon gave up and handled diarrhoea and vomiting simply by sleeping the patient in the bath where the tap was opened the next morning to rinse him down. He was fed the minimum in an effort to counteract diarrhoea and all medication was abandoned. Three months after discharge an emaciated patient was placed in foster care through

social work intervention, after which the unmarried mother deserted him. In other interviews parents would only tacitly admit to the time consuming nature of treating the patient, being careful not to sound complaining especially while relatively good health was maintained.

> 096
> "It's only the physiotherapy three times a day that I find a bit long, but a friend comes round every day to help me".

Adequate control of symptoms went together with the temporary luxury of not facing up to the full implications of the illness. Persistent symptoms on the other hand disturbed this short-lived equilibrium and immediately raised anxiety in mothers.

> 110
> "I know he has an extra nail in his coffin with this cystic fibrosis but I just don't want to think of it".

They were acutely adept at interpreting the seriousness of a cough, the consistency of stools and the complexion or even moods of the patient: (001) "It's like driving a car - you know immediately". The instant response was for mothers to run through their minds what they could possibly have done wrong or omitted either in diet, physiotherapy or medication. In other words they would wrongfully attribute the cause of symptoms to a failure on their part to treat effectively, rather than regarding it as the accepted course of the disease. Mothers tended to see themselves as determinants of both good health and illness of the patient. Two excerpts from one interview further illustrate the crushing effect of persistent symptoms on mothers' emotional well-being.

> 022
> "There are many patients worse than B, she is actually quite well. But to have her like that does not just happen on it's own. You must realise it comes with help from Above, with constant good care, physiotherapy every day, good wholesome food with every meal and looking after her every instant". Later in the interview when the presence of symptoms at night were described, the following was said: "She did not sleep at night, she was so ill and coughing incessantly. I don't think anyone can adequately describe the cough of a cystic fibrosis child - no cough mixture can stop that - I

> think they will even cough under anaesthetic. That cough had me hysterical. There were times when I thought it would be best to end all of it. I started planning when, where and how. I would take her with me in the BMW and we would drive flat out for that one tree which I had selected on the curve of the road - that would end all. But the next morning you feel sick on the stomach with guilt when she smiles at you".

In contrast to the other anomalies, the passage of time did not bring much relief or improvement in symptoms. More often later interviews revealed the feeling that time was running out as symptoms tended to become more threatening.

> 001
> "With Arlene I cannot say, 'wait your turn, when you reach your sister's age you can also have this or that'. I have to do it for her now, because I don't know whether she will live long enough to have what the other children had".

> 110
> "Did you notice how pale B is? The last few nights he has been coughing badly. I am running the humidifier lately to try and help him to get some sleep. I know he is deteriorating but professor D is hiding it from me. I want to know how long I still have with B, because I have got some plans for him to visit the USA where my parents lived and where I was born".

Four mothers of patients (001,022,110,095) who were 10 years and older, revealed that they had consciously crowded the patient's life with activities in an effort to live in the present, because the future was so uncertain.

> 095
> "Look, we knew that she had it in a severe degree. In anything feasible, she participated as long as she possibly could - played tenniset till 9 months before she ... She played the piano beautifully, did ballet and amateur theatre. She also used to say, 'Mum, as long as I live, I must live fully'. Even when she was weak, towards the end, she would unpack and tidy her cupboard, make a dress and cook meals with special seafood dishes".

Increasing age in patients similarly brought increasing difficulty in hiding feelings of sadness from them as disturbing symptoms became more apparent. This was acutely felt by 7 mothers (001, 022, 026, 108, 110, 095, 096).

022
"She is a thoughtful, highly intelligent child. She reads my mood, she reads my eyes. For the last 6 years I have been one helluva actor. I smile outside, but inside I am crying for her. At present it is very difficult at home and I am so tired of wearing a mask and playing the happy role".

096
"You see, I never let E see how I feel. When I am terribly upset I go into my room and cry alone - I don't want her to see what I feel".

Older patients became emotionally resistant to treatment and where parents felt that this was alienating the patient they responded initially by coaxing, only giving up when the terminal stage had been reached, as the following excerpts respectively indicate.

022
"I was shocked when she shouted at me, I did not recognize my own child when she said, 'I have decided, I will rather die than have more treatments! I am finished, I am sick and tired of this business of coughing and medicines and treatments, I wish I'd die!' My reaction was totally wrong because then I started shouting and we both ended up crying".

102
"On the Friday night we knew that he needed physiotherapy, but he got very upset and he wasn't very strong. So on Saturday I also did not give him physiotherapy at all. We didn't do it again before he died, because it was too upsetting for him".

With the older child, the growing feeling of losing a grip on the control of symptoms was accompanied by apprehension of the penetrating questions from patients about the prognosis of cystic fibrosis. At the time of study 5 patients were still too young to understand such information, a further 5 had died at too young an age, leaving 7 patients where this had become an issue. When questioned, the latter mothers all thought that the patients knew the term cystic fibrosis, but only 3 thought that they had known that it was fatal, two of whom both died at the age of 16 years. In the two last mentioned cases the near terminal stage had been reached before the fatal nature of the illness could be discussed. Even then it was rationalized by some as the interview, a month prior to the patient's death, reveals.

108
"I told him all about his sickness, that he has to take his medicine, he must take his treatment. It is so important for me to keep his lungs clear and he doesn't want to anymore. He asked, 'Can I die with this?' I said, 'You can die anyway, anyone can die, just take your treatment, then you will be all right' ".

In the second case the patient had known and accepted death. The one remaining patient knew that cystic fibrosis was a lethal illness, the family having lost a 6 month old sibling as first born. When experiencing a spell of ill health, death was anticipated by this patient.

096
"I told her everything. Earlier this year she was so sick and normally I go to the church service because she likes to have some time alone. But she said to me, 'Mom, please stay with me today' and she asked for communion to be served to her. She also knows that in actual fact children are only loaned to us".

In the four remaining cases, discussing the outcome of cystic fibrosis had been avoided although threatening questions originated at school. At home parents questioned what should be said and tended to postpone facing the issue, but at the same time remained uneasy about it.

001
"I have been honest with her, but there comes a time when you want to know how to tell her that she will ... I believe she's got to know more. She won't find out anything easily, because I had a job finding out about cystic fibrosis. Overseas they have got it published - almost advertising it. Here it is more sheltered. But I mean she will want to know more. Now, must she or mustn't she know that ... I spoke to Mrs M - her daughter is 16 and knows - she said, 'You have got to tell her the truth and then stand by her, you will always be there'. But I don't think it's worthwhile, she's looking so well at the moment, she's also just had her first menstruation, I can almost not believe that she will ... she is such a positive child".

110
"He knows a lot about cystic fibrosis, but I've always kept it from him that it's ... after all, he could be run over by a car tomorrow and who knows anyway when they are going to die?"

Where an older sibling had died of cystic fibrosis, remaining silent about the disease became more complicated. Although parents were at a loss as what to do, advice from "outsiders" was resented, as the following illustrates.

022
"The day will come when she is going to ask me and I don't know what I'll say. What are the right things to say? Should I say that I don't know? Unasked for opinions I've had enough - armchair critics usually have all the answers anyway. The last couple of months she talks a lot about the future - what she wants to do, the number of children she wants to have - and then she looks at me, watches my response. Maybe she's fishing for answers or just using her common sense. Remember she knows her brother died of cystic fibrosis, saw the photographs of the skeleton-like figure. Then they say I should never have told her about him. I should not have mentioned the word cystic fibrosis, but how on earth could I hide that from her? But she knows very little, only the name cystic fibrosis, that's that. She doesn't know that I shudder at the thought of her saying to me straightforwardly, 'Mom, do you think I'll make 20 years?' ".

As the foregoing discussion indicates, parents of patients suffering from cystic fibrosis gradually became technically adept at reading and treating their symptoms. However, long term adjustment was thwarted by increasing severity and the threatening nature of these symptoms. Additional stress was experienced in the parents of patients who were 10 years and older where a growing resistance to treatment developed as well as increasing questioning of the full implications of the illness.

2. Oesophageal atresia

In contrast to the foregoing the difficulties in handling the residual symptoms of oesophageal atresia at home, diminished considerably with increased patient age. Although choking on foodstuffs was frightening, hope of an improvement as the patient grew older, was an important supportive factor to parental attitude as the following indicates.

083
"I remembered the day I read in the hospital folder: 'responsible relative' with my name filled in next to it. There the realization of what I knew already came to me consciously: I would have to see this baby through his problems, there would be no turning back. But at least as each day passed, I knew that he was growing stronger and I had faith and such a sense of well-being, that nothing could get us under - eventually he would be altogether healthy."

At the time following discharge, 19 (83%) of the babies treated for oesophageal atresia experienced serious feeding problems. Of the four remaining babies in this group, two had died before discharge, one had never experienced feeding difficulties and the fourth had shown only mild symptoms which had not been viewed as unpleasant by the parents.

Where feeding problems were present, the feeling was that a situation which should have been pleasurable for both mother and child, could unfortunately be described as "driving any mother instinct left out the window" (095). From the histories it became clear that the establishment of a mutually satisfying mother-child relationship was not only postponed by extended hospitalization but further thwarted at home by this unpleasantness at meal times. In one instance indication of a reinforced death wish for the patient was found.

048
"There were times when I felt, dammit I've had enough now! I asked our GP to come and watch this desperate business of feeding her. I wanted him to see just how this poor little blighter was suffering, trying to feed. Dr X is a marvellous chap, the sort of person one could say this to. I said,'When you take her into hospital again, do me a favour and find a crummy anaesthetist.' I remember thinking afterwards, that I had put it rather crudely. But it was agony feeding this child - your maternal instincts were all screwed up."

Three aspects of residual symptoms were problematic during the early stage of long term adjustment, i.e. anxiety related to frequent choking, repugnance at the "bath of phlegm" during feeding times and the recurrence of chest problems. The intensity with which these problems were experienced, is well illustrated by the following histories.

018
"There was just this increased amount of saliva that seemed to be with this sort of bolus of food in his throat and eventually he could get it out. It used to come out all over the table ...!"

016
"For the first 6½ years she choked on everything - even a grain of rice. We had to watch her constantly. If she remembers to eat slowly and chew well, it's fine. But I was always afraid when she was with friends - they forget".

040
"Just before his second birthday, he was very sick. He kept on getting bronchitis and he had such a high fever, his whole mouth was ulcerated and apparently right down into the trachea. I could not take it any longer and I had to take tablets from my doctor for my nerves, I was absolutely finished. I phoned him and I said look here this child, I can't take it any longer so he said just bring him in. They admitted him straight away and he was there for about 14 days and then before they let him go home they made me see the psychiatrist. From then on it was up to the psychiatrist and social worker whom I am now seeing, whether he could come home or not".

Eight (35%) of parents with oesophageal atresia patients reported recurrent chest problems following discharge of which the effect described above was the most severe. However, evidence of temporary feelings of alienation in mothers to patients during this unpleasant period of coping with symptoms, were found in 43% (10) histories of the oesophageal atresia group as a whole. Feelings of enstrangement within patients towards their parents as described earlier at the time of first discharge, was found to extend beyond the confrontational stage for those patients requiring frequent readmissions. The following history illustrates the nature of these difficulties.

064
"I was petrified because he would lose his breath and by the time we got to the children's hospital he was back to normal again and in the end I phoned through to the ward and I said, look, please I can't handle this child anymore. So they kept him under observation. Well, the way Professor A explained to us the rings of the trachea had softened and collapsed. During that time he was more in than out and of course when he came home we were ready with masks and white uniforms because we were strange to him, he was more used to hospital staff than us".

However, histories did reveal a gradual coping with residual symptoms till they were handled with ease by both parents and patients. As the mother of a 4-year old explained:

> 010
> "Sometimes when he has chicken it still sticks, but I think it is a bit of an act because he can go on swallowing nigger balls and bubble gum without getting into trouble!".

Ample evidence was found in the histories of the patients themselves soon acquiring proficiency in handling residual symptoms (i.e. food bolus) in a socially acceptable way.

The duration of coping with residual symptoms at home

More than half of the parents in the oesophageal atresia group felt that residual symptoms had ceased to be of any consequence by the time patients had reached the age of 4 years (Table 25).

TABLE 25

AGE AT WHICH SYMPTOMS WERE SATISFACTORILY RESOLVED

AGE(YEARS)	N	%	ACC.%
0 - 2	4	17,4	17,4
3 - 4	8	34,8	52,2
5 - 6	3	13,0	65,2
7 - 8	1	4,3	69,5
9 - 10	-	-	69,5
11 - 12	1	4,3	73,9
13 - 14	2	8,7	82,6
Died	2	8,7	-
Unresolved	2	8,7	-
TOTAL	23	99,9	-

As the accumulative percentage in Table 24 indicates, 70% of patients had reached a symptom free stage at 8 years of age. The two patients where symptoms were still being experienced as a problem, were both only

two years of age and can therefore not be regarded as finally unresolved. Table 25 does not include conditions which required orthopaedic treatment (mostly scoliosis) and three patients who were underweight for age, as these symptoms had not been seen as problematic to their parents.

3. Hirschsprung's disease

The unpleasant nature of residual symptoms in Hirschsprung's disease was mentioned as an obstacle to easy assimilation of the patient at home by the majority of parents. Anal dilatations elicited such vehement resistance, that the value of giving such treatment at home may well be seriously questioned. Many mothers confessed to leaving this undone in spite of resultant guilt feelings about the patient's physical well-being. Difficulties related to the technical aspects of colostomies were greatly relieved by contact with stomatherapists where their services had been available. As will be seen later in the discussion, some other aspects still yielded problems. Skin excoriation similarly remained a problem in spite of all the barrier creams offered. Therefore considerable effort went into finding solutions for this problem, ranging from "boererate" to simply leaving patients naked and hosing them down when required.

Eleven (37%) histories described toilet training as an acute point of family argument, especially after the evening meal when most patients had been required to attempt at having a bowel action. Nine patients had questioned why siblings had not been subjected to this punishment. For these families evenings became a time dreaded for its pleading, coaxing and fighting in an attempt to establish some regular routine in

bowel habits as well as a degree of control. Parental efforts varied from creating an intellectual understanding of the physiology by means of simple reading material with illustrations of colonic function to simply rewarding patients for every stool passed.

In Hirschsprung's disease the positive correlation between chronological age of the patient and overcoming residual symptoms, did not follow as clearly outlined as in oesophageal atresia. Amongst others, factors such as severity of the anomaly, the nature of residual symptoms, social class, parental attitudes and patient motivation were simultaneously interactive in accerelating or slowing down the process of adjusting to and handling of residual symptoms. Furthermore the age at which operative procedures were completed varied widely from one patient to the other. As will be shown in the discussion, the eventual outcome of Hirschsprung's disease cannot be ascribed to surgical results alone but should be read together with the abovenamed factors, to name but a few.

The parents of 9 (30%) patients stated that symptoms had resolved to such an extent, that Hirschsprung's disease was no longer of any consequence in the patient's life. A further 14 (46,7%) respondents reported residual symptoms in patients which varied quite extensively, but were nevertheless regarded as minor problems which could be handled. Six parents (20%) were still actively trying to cope with the anomaly, in their own opinion, not always successfully.

Age and severity of illness

In an effort to bring some perspective to the complexity of affecting variables, Schedule 5 depicts the three cagetories described above (i.e. no problems, minor problems, still a problem), together with patient age

SCHEDULE 5

SEVERITY OF RESIDUAL SYMPTOMS BY LENGTH OF BOWEL INVOLVED AND PATIENT AGE

NO PROBLEMS			MINOR PROBLEMS			STILL A PROBLEM		
Research number	Length of bowel	Age	Research number	Length of bowel	Age	Research number	Length of bowel	Age
079	short	2	076	short	1	082	short	7
056	short	4	061	short	1	099	long	10
008	short	6	044	long	2	100	-	15
034	short	7	075	short	2	003	short	16
059	short	13	036	long	3	038	long	19
025	short	14	078	short	5	037	short	21
043	short	18	002	long	6			
087	short	19	074	long	7			
109	short	8	063	long	8			
			035	short	9			
			005	short	9			
			042	short	9			
			060	short	11			
			080	short	20			

and the length of bowel involved. This data indicates that age as a single factor showed a very wide scatter in all categories of adjustment to residual symptoms. As both parental and surgical view of success hinged largely on the degree of continence reached, movement to a better category can still be anticipated for a number of patients, markedly so for those of 5 years and younger (see Chapter 3). Six such patients are depicted in schedule 5 as experiencing minor difficulties. Furthermore as the discussion will show, other factors than age have played a role in acquiring continence even at a much later age.

As expected the severity of the anomaly as determined by the length of bowel involved seems to have exerted some influence on residual symptoms. All respondents with no problems were diagnosed with short segment Hirschsprung's disease (Schedule 5). This data is further summarized in Table 26 which reveals that 42,8% of all short segment Hirschsprung's disease patients had been rated by their parents as experiencing no problems, while this was not true of any patients with a long segment of bowel involved.

TABLE 26
RESIDUAL SYMPTOMS (PARENTS RATING) BY LENGTH OF BOWEL AFFECTED: HIRSCHSPRUNG'S DISEASE

Rating	Short segment %	Long segment %
No problems	42,8	-
Minor problems	42,8	62,5
Still a problem	14,3	25,0
Died	-	12,5
TOTAL	99,9	100,0

The latter tended to fall in the categories of minor to serious problems. A large grey area including patients with both long and short segment Hirschsprung's disease exists in the category with minor

problems. At this point a closer look at the histories is indicated with the view to further clarifying the basis of parents' ratings, especially in this socalled grey area.

Patients with no problems

Parents of 5 of the 9 patients with no problems (008,025,079,056,034) could not recall when continence had been reached, as this "probably fell within the normal limits". Post-operatively these patients had been raised "as a normal child" without expecting or experiencing serious problems. Nothing special had been done at home to assist their becoming symptom free. A further 3 patients (059,087,109) reached continence and a near symptom free stage at respectively 18 months, 2 years and 4½ years of age. As excerpts from their histories reveal, parental attitude varied from unconcern to investment of considerable effort to overcome symptoms.

> 109
> "It took a lot of scheming and trial and error to really find out what he could eat. When he had diarrhoea or this stomach block, I would sit down and think what this child could have eaten to cause this - and I know everything he ate. So he was in nappies till 4½ years. At present it means only watching his diet. He is terribly good, never has an accident in his underpants. He is completely recovered."

> 087
> "Almost immediately following his discharge I went back to work. The maid handled his symptoms, which disappeared after 2 years. He is a perfectly normal child, playing rugby as well as soccer."

The last patient in this group was rated by her mother as having no problems, was married and at the same time successfully holding down a job.

> 043
> "She is totally independent of anyone. She still avoids certain foodstuffs as it might make her run, but really is fully recovered."

In this group a mutual factor at the time of study had been the exercise of some care in the choice of foodstuffs. This was more or less adhered to by patients themselves who were otherwise leading a normal life, unaffected by Hirschsprung's disease.

In two cases a specific incident in the life of the patient had been identified by the parents as the turning point towards this level of functioning. From this time onwards patients also seemed to have taken responsibility for their own well-being.

> 059
> "I was so much looking forward to that moment when she would be without the colostomy. Sometimes I feared that she would go around like that for the rest of her life. Then the operation was done and I almost wished the time away for the day that she would be continent and able to wear panties. I then decided that one had to do a thing which the child could understand and fully comprehend. I took her to the shop where we bought her a number of beautiful panties. At home she chose the nicest one to put on immediately. I then explained that such lovely clothes could not be soiled. Now she would really have to run for the toilet very quickly when necessary. And it worked! She looked after them - she was continent."

> 109
> "I used to buy those towling pants and let him wear them under his proper pants or pyjamas - day and night, rather than wash the soiled sheets and pants of a near 5 year old every day. Then one day he came and showed me his pants. 'Look here, mom, this is clean, why do I have to wear them - John doesn't wear them?' "

Patients experiencing minor problems

A specific incident of the nature described above was mentioned by the parents of one patient in the group of 14 who was still experiencing some difficulties with residual symptoms. This patient with long segment Hirschsprung's disease was one of the most severely affected and had experienced a number of physical complications requiring lengthy periods of hospitalization. Yet the history reveals the following favourable report.

002
"When she was smaller, she used to mess every night and often during the day - there was no control of faeces. Prof. said that by the time she was a teenager, 13 or so, she would physically be better equipped to gain control. On her 5th birthday the most amazing thing happened. That morning she came to us and said, 'Mom and Dad, I am a big girl now. From today I am not going to mess anymore'. And she didn't ! This astounded us - we could actually take her out for a whole day. Since then she has been clean."

The reason why this patient had not been rated as problem free by her parents was the fact that unhappiness had originated at school due to her explosive and malodorous stools which had given rise to teasing and some ostracism. Emotionally the patient had not yet been able to deal with this.

The discussion so far indicates firstly that various considerations influence the rating of residual symptoms by parents. Secondly these ratings could be influenced by factors other than purely physical reasons. One such reason was at least partly centred in the patient's conscious and active commitment to overcoming residual symptoms and taking responsibility for their own well-being. Thirdly where difficulties had been experienced, they could not always be directly ascribed to the anomaly itself but should more accurately be termed as secondary maladjustment. For example in 002 cited above, adjustment to a still better level could be achieved without any further improvement in symptoms per se. However, the realization that more subtle handling of such symptoms (i.e. offensive stools) could eliminate the source of secondary maladjustment (i.e. ridicule) had not yet grown in this young patient who had already done so well.

Other histories confirm the unnecessary limiting effect of so-called sources of secondary maladjustment. For instance, judged purely on

symptoms, a patient's long term results were excellent as described by his mother.

> 035
> "From the age of 18 months he was out of nappies and fully continent. We never forced him, but he never had problems. He was 100% clean, even at night. Now at the age of 9 I find that he is very fastidious with underpants - probably because he realizes that he is slightly different as far as bowel habits go".

Despite these excellent results the parents rated the patient as having residual difficulties due to Hirschsprung's disease. This hinged purely on the attitude of the patient's current class teacher. During the years when a good relationship had been established, he was permitted to sit in the front desk and nip out to the toilet without specially asking for permission on each occasion. Noisy and frequent stools then passed relatively unnoticed, facilitating normal adjustment. However, where teachers insisted on a routine toilet break for the class as a whole, adjustment was jeopardized, as was the case at the time of interview with the above patient. These aspects pertaining to school adjustment will be more extensively discussed in Chapter VIII.

When looking at individual cases, social class seems to have had some measure of influence on parental views of patient adjustment to residual symptoms. Two examples can be cited where parents in social class IV rated patients (074,078) as having minor difficulties, even though symptoms could have been seen as offensive. As the following indicates however, symptoms were tolerated with relative unconcern.

> 074
> " Perhaps he'll come home from school today and is clean. Tomorrow he will come and there will be something in his underpants. I warn him to go to the toilet before going to bed but he lies to me and then his stomach will work in bed at night. But I have got a plastic bag over the mattress and not just a sheet. But he is a healthy child and eats anything we eat."

078
"He does not seem to have much control. Sometimes he also soils himself at night. This can happen every night for a week in a row and then disappear again. We have been wanting to take him back for a check-up, but havent got round to it."

Patients experiencing Hirschsprung's disease as a problem

In contrast to the foregoing, exactly the same symptoms as above were described as major problems by two parents in social class II and the patients were rated as still experiencing Hirschsprung's disease as a problem (038, 082). In one instance this was verbalised as follows.

038
"After 18 years we are still battling with Hirschsprung's disease - it is a problem. He copes marvelously by himself during the day, but as soon as he sleeps, everything just runs. So it's a question of nappies and drawsheets every night. Look, we have to live with it ..."

Although no direct relationship between social class and severity rating can be claimed, histories seem to indicate that social class factors do contribute in part toward parents' rating of end results.

The excerpts of histories discussed earlier, have sufficiently illustrated that in some cases positive patient attitudes had been conducive to favourable or at least more acceptable outcomes for Hirschsprung's disease. The following history illustrates the reverse attitude where a patient seemed to have had no interest in outcome. At the same time the socially destructive nature of residual symptoms in Hirschsprung's disease is shown, both within and outside the family circle of the patient.

003
"She is coming along very nicely but the only thing that I have got to moan about is that she is still careless about hygiene. Her bag is practically due for a change but ag, she'll do it tomorrow. She'll go out in the afternoon and the next minute we have the odd winds coming out, you know, the smell and I can just see red because there is no reason for it ! There is no reason why she must smell at all. Another thing I get very annoyed with is that

> she knows that certain foods upset her, but she will go and eat them and within a few hours, right in company (I don't mind here at home so much), these smelly whiffs come along. Look, I can still take it but she herself, she is very aware that she's let off and it is smelly. She cringes every time, it is for her sake that I get annoyed about this. I feel if I don't see to it now that she is a teenager, she will slowly get more sloppy and it will get worse and worse. Just now she ostracises herself completely from people. Basically she does not care, she's got that attitude I don't care and she can let herself go. Yet if I do too much she is up in arms and defiance. That's the only problem I've got with her. I picked her out on Saturday and I really went for it. She got cross too - said 'I suppose you could not wait to get in the car to pick me out' and I said 'you are right, I could not wait ! You are now nearly 17 and you should know that you are going to a party this afternoon where you are going to mix with ladies. You should not have had the sweets and what have you in the morning because you know that it is going to cause winds in a few hours. Rather wait until you've got nothing on socially. I don't mind if you eat anything, eat all the soup in the world but do it at night or do it when you are not going to mix with other people, discipline yourself.' Well, we are still friends. We both go our own ways for a couple of hours, or sleep on it and the next morning you know, it's all over fortunately."

The reason for experiencing Hirschsprung's disease as a major problem, was sometimes related to the patient's own particular set of circumstances. For instance, where intermittent soiling had been satisfactorily handled at home by the patient cleaning up both himself and his underclothes, the necessity of having to attend boarding school as the only available education facility, in one case gave rise to considerable worry. Especially night soiling was feared as this would elicit ridicule and possibly ostracism. The parents rated this patient as experiencing Hirschsprung's disease as a major problem.

Where dependence on the mother for remaining clean had not yet ceased at a relatively late age, Hirschsprung's disease remained a problem. Two mothers of a male and female patient (038,037) of respectively 18 and 20 years of age daily attended to them in this respect at the time of study. Both mothers were apprehensive of the day when "I won't be around to do so any more," yet did not see their way clear to leave this up to the patient. However, for the Hirschsprung's disease group as a

whole, mothers more often referred to their reticence to talk about bowel habits as the child became older. For some the only knowledge of residual symptoms was the odd piece of stained underclothing found in the wash or dustbin. Many had reached an unspoken agreement that stained underclothes would be the patient's own responsibility and not part of the family laundry. Some mothers admitted to actively distancing themselves from involvement with residual symptoms as this became progressively embarassing, especially between mother and son. It seemed that, although such mothers had remained uneasy about residual symptoms, patients did manage to obtain their permission to sleep over with friends or attend a week-end camp. This was often the opportunity when patients for the first time verbalized their own doubts about "coping with my bowel on my own". Although parents were unclear of how this was in fact handled, there was consensus that more self-confidence and a sense of achievement ensued for the patient. It was concluded that parent attitude had made some contribution towards healthier adjustment in spite of socially limiting residual symptoms.

During interviews the impression was gained that some parents might have underestimated the serious nature of Hirschsprung's disease and also had too high an expectation of surgery alone as a determinant for a symptom free result. This became apparent, firstly by expression of an oversimplified view of the procedure as removing "sick" bowel to leave only healthy function. Secondly, where for instance a patient (087) had reached full continence by the age of two years, both parents regarded this as rather a later recovery. Similarly where a patient (025) had never experienced any residual symptoms, this was merely seen as a normal expected result. Thirdly, some mothers had been under the impression that the residual symptoms in their child had been more or

less an isolated case of a poor result. Whether the erroneous oversimplification originated from surgeons' explanations or from parents misunderstanding, was not possible to determine.

4. Anorectal Malformations

As in Hirschsprung's disease, the very similar and unpleasant nature of residual symptoms in anorectal malformations counteracted easy assimilation of the patient at home. Again anal dilatations were loathed and much relief experienced when they were no longer necessary. At the time of study 15% of parents felt that the anorectal malformation yielded no problems worth noting. Eight parents (40%) were experiencing some problems with residual symptoms in their child, but were managing. A further 40% of parents still saw the anomaly as a problem. One patient who died as a neonate, was not included in the present discussion.

In Schedule 6 the age and anomaly classification of the above three groups are depicted. This reveals that 3 of the 8 patients with minor problems and 5 of the 8 patients where the anomaly was still a problem were 5 years or younger. As the age between 2 and 5 years requires major co-operation from parents in motivation for mastering continence and at the same time preventing constipation in the patient (Chapter 3), movement to a better level of functioning is highly likely for an additional 40% of the anorectal malformation group. As individual histories will show, this is also possible for the remainder of patients who were much older.

In contrast to Hirschsprung's disease, the severity of the anomaly, (i.e. betwen high and low anomalies) seemed to exercise very little influence on the adjustment made to residual symptoms (Table 27). In

SCHEDULE 6

SEVERITY OF RESIDUAL SYMPTOMS, HIGH AND LOW ANOMALIES BY PATIENT AGE

NO PROBLEMS			MINOR PROBLEMS			STILL A PROBLEM		
Research number	High/low anomaly	Age	Research number	High/low anomaly	Age	Research number	High/low anomaly	Age
045	high	17	007	high	18	027	high	11
049	low	3	024	high	11	050	low	2
103	low	9	032	high	12	066	high	3
			033	high	7	067	low	1
			057	low	4	084	low	2
			062	high	3	088	high	6
			070	low	5	092	low	5
			090	high	13	094	high	14

order to find some explanation for this discrepancy, a closer look at individual histories is required.

TABLE 27

RESIDUAL SYMPTOMS (PARENTS RATING) OF HIGH AND LOW ANORECTAL MALFORMATIONS

RATING	LOW ANOMALY %	HIGH ANOMALY %
No problems	25	8,3
Minor problems	25	50,0
Still a problems	50	33,3
Died	-	8,3
TOTAL	100	99,9

Patients with no problems

In the first instance having no problems was attributed by the mother to the success of surgery, with the residual constipation well handled by both mother and patient.

> 049
> "He just moved her anus back where it belonged and it's fantastic. She looks perfectly normal down there. She has to have laxatives every day but the problem was discovered at birth and treated."

In the second instance where a very lengthy and difficult pre-diagnostic phase had been experienced, diagnosis brought much relief. Over time some distancing from residual symptoms by the mother occurred.

> 103
> "She does not ever complain anymore - but then she is at school now with such a lot of other things happening, (taking her attention). Physically she seems fine, not underweight, not undersized. I think she would tell me if she was not coping with her stools. Maybe I had better ask but little girls get fastidious about things like that ... ".

The only patient (045) with a high anomaly having reached good adjustment to residual symptoms, achieved this level at the age of 16 years. His mother described a specific incident which marked this change. In the past he had not watched his diet, soiled himself and had been ostracised at school for being smelly. Mrs E. largely blamed

teachers for not permitting him to go to the toilet freely. In the second term of std. 9, the patient "gave up the struggle" and refused further school attendance. When signing up as a policeman, he explained the necessity of frequent toilet habits to his superiors who accepted this. Since then personal hygiene, diet and continence had ceased to be problematic.

> 045
> "He seems to be very happy in his work and his personal hygiene has changed overnight. I don't have to remind him to avoid orange juice or not to have mayonnaise on his salad. For the first time in his life he watches his own diet and what's more you can't lay a finger on his personal appearance."

In the above history the mother added that she felt freer to move somewhat away from her son in a relationship which she described as warm, but perhaps too close and protective for a son of his age.

Patients experiencing minor problems

The type of problem experienced in this group is well summarized as follows and more or less reflects the status quo of 7 patients.

> 070
> "He still blocks up sometimes and has to take medicine for this. Although he has trained himself well he still has accidents. If he takes opening medicine, it comes out with a rush and he can't cope with it. Like any child playing, he gets distracted and then they don't want to go."

In all instances mothers felt that they had been unable to obtain full co-operation from the patients themselves. Three histories revealed some concern in mothers that the relationship with their sons, respectively 11, 12 and 18 years of age, had had to include discussion of continence. It seemed that fathers had been largely uninvolved.

The one remaining patient (090) aged 13, with the most severe type of all the anorectal malformations, cared meticulously for her own

colostomy and refused to be restricted in activities to the extent of also participating in swimming. Her difficulties revolved mainly around impatience to have further plastic surgery done to obtain a cosmetically more acceptable result for her genitalia. Changing into sportswear at school had elicited unwelcome comments from pupils about the anomaly.

Patients experiencing anorectal malformations as a problem

Symptoms in the 8 patients in this group influenced family life to a greater extent than those discussed above, as the following history reveals.

> 092
> "For me this is still a major problem. She wears a nappy every night and because of this I think, has become lazy with toilet habits. I have resigned myself to this. At a stage I tried to help her by waking her every three hours. Later my husband complained about the disturbed sleep and said 'Just put a nappy on and leave her'. I know we also give in because we are sorry for her - she has always gotten away with more".

Again lack of co-operation from patients was commonly seen as an obstacle to controlling symptoms. In this group one mother expressed a distinct dislike to dealing with her son's symptoms. Her feeling was that their relationship had been negatively coloured by the anomaly to such an extent that both her husband and friends had warned her about this. At the same time guilt feelings about not assisting him in successfully gaining continence were expressed.

In one instance (050) cosmetic effect (in a diagnosis of vestibular anus) served as main motivation for a parents rating of the anomaly as a problem. Surgical refusal of further operations to moving the anus further back was based on the high risk of creating incontinence in the patient. However, the father insisted that marriage could not be envisaged for his daughter with the present results.

B. COMMUNITY REACTION AND PARENT ATTITUDE

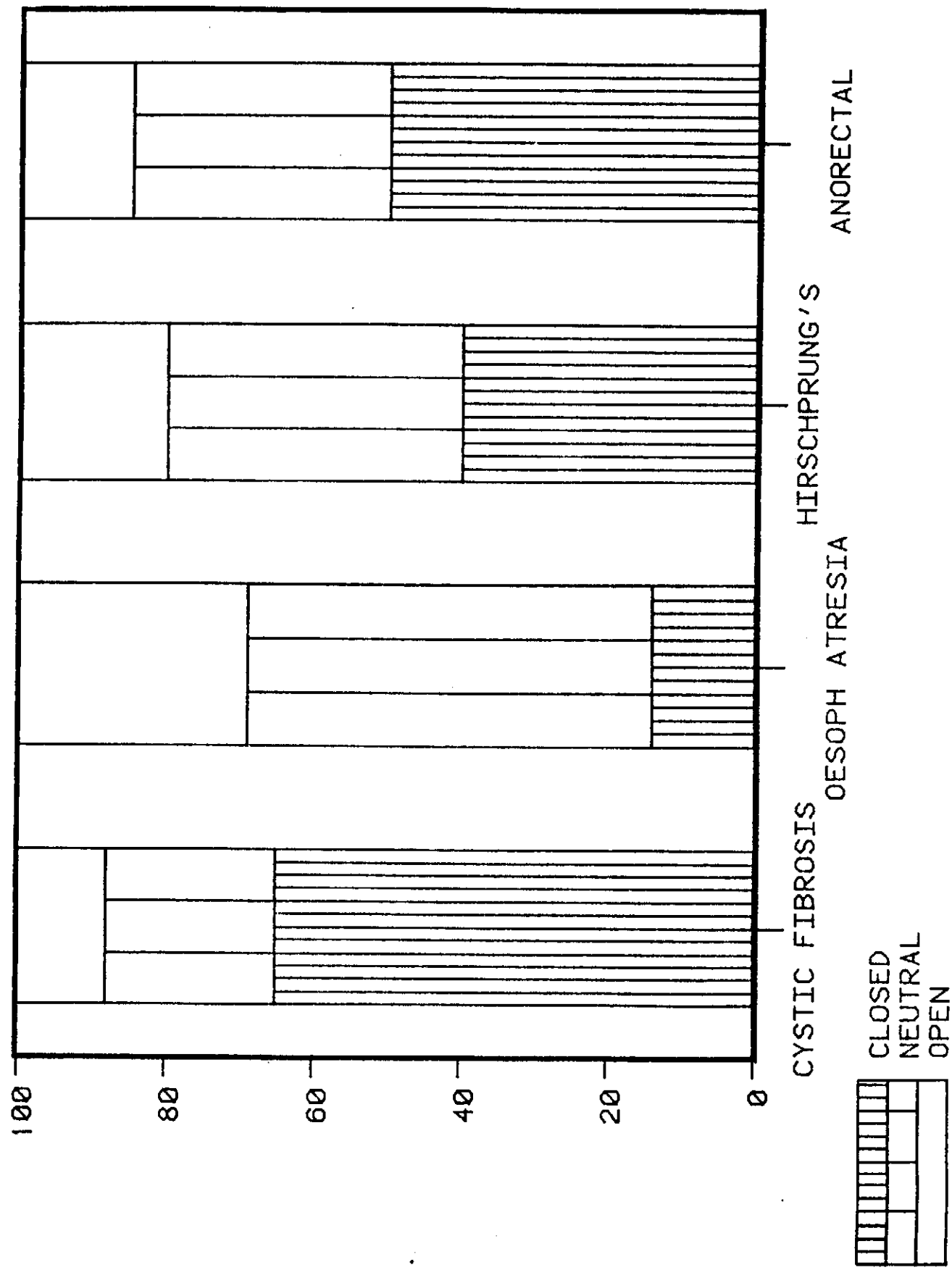
During the course of interviews community reaction was often raised by parents either as a limiting or as a helping element in dealing with a child with a congenital anomaly. When extracted from the histories, parental attitude towards community reactions could be classified into three broad categories. Firstly there were parents with an open attitude towards the community mainly because they had experienced the benifits of its support. Secondly, there were those who had closed family ranks towards the community either because of shame or after having experienced negative attitudes from the community. This left a last group where neither of the above could be detected in the histories and were for summary reasons therefore classified as neutral. Although these attitudes are discussed as they presented at the time of study, clear evidence existed that there had been some change over time. Furthermore although the reason for such attitudes differed between the anomaly groups, the overall indications were that families had benefited from open attitudes - though not in outcome but certainly in handling difficulties with greater ease. Figure 2 depicts the proportion of histories reflecting open and closed attitudes towards the community.

1. Cystic fibrosis

The parents of 11(64,7%) patients with cystic fibrosis described a closed attitude towards the community. In all instances the typical reaction had been that of "people don't know about cystic fibrosis, nor do they understand when it's explained and therefore it does no good to discuss anything with them"(001). The most severe overt reaction (012) was illustrated by the mother who became so isolated from the community

FIGURE 2

PROPORTION OF HISTORIES REFLECTING OPEN OR CLOSED ATTITUDES TOWARDS THE COMMUNITY



that she was unable to leave the home, nor was she prepared to receive visists from anyone. She attached herself to another parent of a cystic fibrosis patient, who in turn referred her to the social work department as the relationship had become too demanding for her.

Even in homes where cystic fibrosis was otherwise well handled, the absence of community understanding was felt.

> 110
> "People don't understand cystic fibrosis, so I just quietly avoid discussing it. We used to have neighbours who took the trouble to really investigate what cystic fibrosis is all about and got to know it's serious nature. They have moved and now I feel quite cut off again."

In some instances parents thought that they were not believed or that the reaction towards the patient was wrong, which separated them from the community as the following two histories respectively illustrate.

> 081
> "When you do explain cystic fibrosis they don't believe you. They look at your child and say, 'but she looks so healthy' and you realize they don't believe a word."

> 108
> "Because of cystic fibrosis he had a very small stature. No one would believe that he was 16 and even adults treated him like a child of 5 years. He suffered a lot because of their attitude."

Due to the difficulties of explaining cystic fibrosis to laymen, some parents had summarily rejected any friendly questioning as curiosity and refused all sympathy as the following histories indicate.

> 022
> "They mustn't even enquire after her well-being because I don't want to talk about it. It's only curiosity and how can I tell them not to talk about cystic fibrosis and just to leave me alone? I don't want to see sympathy in anybody's eyes ! Must I lie to the world and say she's fine?"

> 071
> "I don't go anywhere, not because of her, it's just that I don't want to talk about cystic fibrosis because it's a small town. Now I have got it down to telling them it's just a digestive problem and that fixes up their curiosity and it does not tell them a lie and then they won't keep on and on."

In contrast to the foregoing, two families both living in small communities described excellent support only, as the one history shows.

> 095
> "She was so well accepted and loved, that everyone worked together towards her recovery. She was always treated as perfectly normal and participated in everything possible till the day she died."

In the second instance (072) the community consisted of a few hundred people in a secluded defence force camp. Here the community was mentioned as the main source of support in raising a child with cystic fibrosis.

2. Oesophageal atresia

Of the anomaly groups studied, the histories of oesophageal atresia patients seemed to reveal the least involvement with community response with 56,6% of parents being classified as neutral. This can be explained in terms of both the limited duration and the more socially acceptable nature of residual symptoms.

> 091
> "It was such a frustration to try and get through to them that he could not swallow normally. They tell you what you should do and that you are overprotective. They could not understand that the oesophagus did not have normal reflexes. This frustration I had till he was about 2, then things improved."

The three (13%) families who had closed ranks towards the community included one patient (093) whose diet was still being watched, a fact which had been criticised by neighbours and friends. The two remaining patients (014,077), had died respectively one year and 4 years before with severe associated anomalies which the parents had been too ashamed to discuss with anyone. Any questioning was acutely resented.

An open attitude was projected in 7 (30,4%) of the histories of

oesophageal atresia patients. As the two examples show, attention had been welcomed and appreciated by parents.

> 052
> "He is history in this vicinity, being born like that. He goes all over with anyone - I think he must be the most well known and well loved baby in this whole place."

> 064
> "All the churches were praying for him and wherever we went, people I don't even know asked how he was. I never lacked for any help if I wanted to go somewhere or do something - someone would always come in and take over."

3. Hirschsprung's disease

In the Hirschsprung's disease group, 12 (40%) histories reveal a closed attitude towards the community. In all cases this was related either to colostomies or residual symptoms with their high possibility of social embarassment. In one family where two children with Hirschsprung's disease were born, the histories clearly reveal an initially closed attitude with it's concurrent hardship. With the birth of the second affected child, moving to an open attitude had brought about easier adjustment. Patients were respectively 9 and 3 years of age.

> 035 and 036
> "I was desperately afraid that he would smell. When visitors came I would peep to see who they were before letting them in - and I wanted no sympathy. I wanted no one else involved. This was a thing that the two of us, (my husband and I) had to fight alone. And my kindergarten children wanted to come and see their teacher's new baby. This was too much for me ! What if they noticed something wrong? With the second one we had moved into a house and had beautiful neighbours. Everyone helped. I showed them how to clean the colostomy and could leave him with anyone of them when doing my shopping. Perhaps there was some meaning after all in going through all of this for a second time."

In the Hirschsprung's disease group it seemed that the necessity for a closed attitude was more acutely felt in the first stages of long term adjustment when equilibrium had not yet been reached at home. The following excerpt again refers to a family with two Hirschsprung's disease patients, in this instance both had been 3 years and younger. Referring to the eldest, the father commented:

075 and 076

> "When people came along and asked questions, it was something we could not handle. Every time I felt that my wife had just started accepting the child and that we were now going to put our heads down and see what we can do to cope with this child, then someone comes with questions and we're back to square one again".
> The mother continued:
> "I was not advertising that they had something wrong with them. But just when I started feeling they were normal, someone would ask about the operation. I tried to act as normal as possible, because they are normal children, they just had a couple of mishaps".

The comments by the father of a 7 year old patient seemed to indicate that although symptoms were still embarassing, he expected to find some degree of acceptance from the community.

> 080
> "Because he can't squeeze one is sometimes sorely embarrassed. You're not prepared, you don't have the right words to apologize to the people around you ... one is constantly alert, tense. On the other hand we're only a small country community, I am sure people must have some understanding."

One history (003) revealed how a small road camp community in their own way actively intervened with the difficulties of Hirschsprung's disease. When realized that a mother was becoming depressed, their efforts amongst others included playing bridge for 12 hours at a stretch to keep her socially involved. Sixteen years later this community support was recalled with much appreciation. This open attitude was repeated in the same family when a second affected child was born 2 years later and died soon after. Whereas all other histories of the surgically correctable anomalies revealed a closed attitude where patients had died, this history was the only exception.

In the Hirschsprung's disease group two examples (078 and 074) were found where parents had always been open to the community and remained so even after negative response to the patients' colostomies. While holidaying in a caravan camp an unpleasant incident in the ablution block was simply shrugged off as "funny". Later in the interview the

mother concluded with the following.

> 078
> "There was an article in the Fair Lady of a child who had a colostomy. This family described how people stared, how they lost friends and were isolated. I found that strange, because we did not experience anything like that".

In the second example (074) the patient of 7 years old was accused of stealing as a shopkeeper mistook the bulge of his colostomy for stolen goods hidden under his jersey. The incident ended in embarassment when the colostomy bag was pulled free by the shopkeeper. Although the mother expressed being momentarily antagonistic when this was reported at home, an attitude of relative unconcern about residual symptoms and the effects was nevertheless found at the time of interview. In both instances the meaning attached to the events by families had not been serious enough to result in a closed attitude towards the community. Overall six (20%) families of Hirschsprung's disease patients reflected a socalled open attitude.

4. Anorectal malformations

Ten (50%) histories of anorectal malformation patients revealed a closed attitude towards the community with regard to the anomaly. Again where a patient died of multiple associated anomalies, interest and questions from others were seen as curiosity and resented.

> 013
> "Not that it's anything to be ashamed of, but we just felt that it was not necessary to tell everybody. The baby passed away and that was that. We socialize quite a lot and were apprehensive about having to answer questions - they're just inquisitive for the sake of being inquisitive."

Two examples (057 and 062) were found where a closed attitude from parents originated for the opposite reason. In other words, there had been a real need to talk to people about the patient's anomaly. However, these parents felt that not enough interest was shown as the following excerpt indicates.

066
"I found with most people when you want to talk, they are not interested, not really. When you are busy chatting they will overrule the conversation by talking about something else to try and avoid this subject - to try and get it off your mind. Meantime you wanted to get it out. Many times it frustrates me terribly and I get very vindictive. I want them to know about my problem, but then I thought they could all go to hell."

Parents who had taken an open attitude (15%) invariably described this as a positive element in raising a child with a congenital anomaly, even though everyone in the community had not always found it easy to tolerate.

070
"When I brought him home after the colostomy, I explained to friends who then asked to see it. They watched while I changed him and one girl's husband got sick - it revolted him completely. I also had the maid circulate the Xhosa literature on colostomy care all around the staff on the farm. So everyone knew exactly what he had and what was going on, so there was no staring at him. We even had episodes where he was cross with his sister and ripped the bag off and chased her with it."

032
"I think in a small town one's friends are much closer to one. And it is a lot of support if you have people who obviously care and are there for the sensation."

In the last instance (033) a family also persisted with an open attitude although earlier experience revealed some negative reaction: while the patient had still been colostomized, he was allowed to play naked in the backyard in order to counteract skin excoriation. When she went out to hose him down, the mother was approached by shocked neighbours with a request to stop this practice which they found unacceptable in the well-to-do neighbourhood where they lived.

The discussion has shown that support, or at least acceptance from the community, had been a felt need for the majority of parents, whether or not an open attitude had been followed. It seemed that initially some time was needed for parents to come to grips with the anomaly, before a readiness for community interest could develop.

Where patients died a closed attitude had been prevalent. In other instances lack of insight from the community as well as parents regarding negative feed-back as a permanent rebuff, had often terminated in a closed attitude. Some examples of perseverance with an open attitude, in spite of negative incidents, were shown. In such cases the meaning attached to these incidents had not been serious enough to change parental attitude.

C. CONTACT WITH PARENTS OF OTHER PATIENTS

In view of the difficulties experienced with residual symptoms at home, it was anticipated that contact with parents of patients with similar conditions would serve as a valuable source of support. For the parents of 35% of the patients in the sample as a whole, such contact had been arranged at one time or another during the course of hospitalization or when attending clinics (Table 28). Only 3% of parents pursued contact for a long enough period to have experienced any benefit. Sixty-four percent had never established contact with other parents. The parents of cystic fibrosis patients had most frequently (82%) been in contact with other parents but again had not pursued such contact for any length of time. In the surgically correctable anomalies, it seemed that such contact was not even tentatively made (70% - 87%).

Table 28

CONTACT WITH PARENTS OF OTHER PATIENTS WITH SIMILAR CONDITIONS(%)

Contact with other parents	Cystic fibrosis %	Oesophageal atresia %	Hirschsprung's disease %	Anorectal malform %	All cases %
Contact arranged but not pursued	76	9	30	25	32
Continued contact	6	4	0	5	3
No contact made	18	87	70	70	64
TOTAL	100	100	100	100	99,9

When parents were questioned about their attitude towards contact with other parents, only 14% saw this positively as a potential source of support (Table 29). The majority of positive attitudes especially in the group with surgically correctable anomalies were verbalized by parents who had already found their feet and could see themselves in a helping role, sharing their experience to the benefit of other parents who were not coping well.

Table 29

ATTITUDE TOWARDS CONTACT WITH OTHER PARENTS

Attitude	Cystic fibrosis %	Oesophageal atresia %	Hirschsprung's disease %	Anorectal malformation %	All cases %
Positive	18	17	3	25	14
Neutral	12	70	47	50	47
Negative	70	13	50	25	39
TOTAL	100	100	100	100	100

There had been an unwillingness to receive help from other parents

although many nevertheless expressed an interest to know "how others were making out." In contrast, parents of cystic fibrosis patients had most often been negative, with the small proportion of positive ones preferring to associate with the parents of patients who were in relatively good health. The obstacles raised by the parents of cystic fibrosis patients were that they were barely coping themselves (023,107,095) and that seeing other patients suffer or die eroded their own confidence (096,102), or that they didn't want anyone becoming dependant on them (012). Even a mother (001) who was positive herself about mutual support, complained of the negative response encountered with other parents. Part of this negativism was further pin-pointed as originating from criticism amongst parents of the way in which other parents handled the child with cystic fibrosis (021,022,071,096,110). The above mentioned obstacles were raised as equally applicable to the Cystic Fibrosis Association, for which parents showed very little enthusiasm as a means of finding support. In addition it was criticized as being run by men, while the major burden of cystic fibrosis was carried by mothers (110), with the result that it was business orientated and not geared to support. Parents who had lost children with cystic fibrosis all felt that they had best avoid the Association, as their experiences only served to dampen what hope other parents still had. Others merely rejected such formal groups without offering reasons: "I was dead against it from the beginning" (023).

The parents of patients with surgical correctable anomalies raised the vast differences in residual symptoms as main obstacle to beneficial contact between parents: "Problems differ too much" (006,027,094,103, 059,060,099). Further objections included that information offered by other parents tended to confuse more than clarify (061) or complaints

that their privacy was being invaded (094) or that they were hindered by differences in social status of the family referred to them (103).

The large proportion of attitudes classified as neutral (Table 28) was again not well substantiated. For example replies were verbalized as follows: "I didn't really need a society or anyone" (003) or "I never missed anything like that" (044).

In reviewing the histories, the impression was gained that a potentially valuable source of support had been lost due to the obstacles mentioned earlier. More care exercised when doing referrals between parents could well have circumvented many of the problems discussed above.

DISCUSSION

A. Handling symptoms at home

It has long been known that treatment results which have been regarded as good by hospital staff have been accepted with reluctance and far less enthusiasm by parents (Woolam, 1964). To a certain extent this was found to be true in the present sample. Parental view of outcomes, on the other hand, did not necessarily correspond with the severity of residual symptoms. Other variables contributed towards the formulating of parents' final conclusions of the measure of success achieved with the outcome of the anomaly.

Cystic fibrosis

1. Whereas other researchers have verified the demanding and time-consuming nature of handling cystic fibrosis symptoms at home (Burton 1975, Falkman 1977), the parents in the present study did not

admit to experiencing similar hardships. Where, however, patients had died, parents in retrospect confirmed the finding of the above researchers. A possible explanation for this discrepancy might be in their "healthy denial" of inevitable circumstances (Ross 1978).

2. The handling of cystic fibrosis symptoms at home was found to be particularly upsetting to parents because of their reminder of the patient's eventual death. The difficulty they experienced in discussing the prognosis with the patient has also been noted by others (Mikkelsen, 1978).

3. Similarly the increasing problem of hiding their own feelings of sadness as the patient became older which was found in the present sample, corresponds with the findings of McCollum and Schwartz (1972). This lack of open communication between parents and patients about the prognosis was also recorded in the study of congenital heart disease (Garson et al, 1978) and of childhood cancer (O'Mally, 1979).

4. The underlying difference in the long term adaptational phase between cystic fibrosis and the surgically correctable conditions was an overriding fear of the patient's eventual death, a fear also noted by others (Driscoll and Lubin 1972, Wagner and Hein 1976). This fear increased as symptoms became less amenable to treatment.

5. Patients' questions about death were poorly handled because parents themselves had difficulty in facing this issue. When discussion did take place, this was most often postponed to the near terminal phase (Kulczycki et al 1969).

6. In further contrast, an increase in patient age was not accompanied by better mastery of symptoms or the hope that time would bring improvement. It was found that the emotional difficulties mentioned above were intensified where patients were ten years or older, a finding which was confirmed by others (Tropauer et al, 1970).

7. In addition, the older patients became increasingly opposed to adhering to daily treatment regimes.

8. Parents on the other hand seemed to over estimate the benefits of their home treatment . This treatment was often only relinquished in the terminal stage.

9. A further reaction peculiar to the cystic fibrosis group was the almost agitated cramming of activities into the patient's life in an attempt to compensate for their limited life span.

Oesophagel atresia

In contrast to the foregoing, the passing of time brought increasing hope of further improvement in oesophageal atresia patients. In the long term adaptational phase the following was found to be relevant:

1. Hope of complete recovery over time was a realistic expectation for this group and was seen as such by parents. This was borne out in the findings that by the age of 4 years, more than half of these patients no longer suffered from any residual symptoms.

2. Residual symptoms mostly caused unpleasantness at meal times due to choking and the development of food bolus when eating. Although symptoms were acute and frightening, patients and parents soon managed to cope with these in an acceptable way. Similar results were recorded by Dera et al, 1980.

3. The nature of symptoms did not cause any embarrassment for parents or social ostracism of the patient as was the case in patients with Hirschsprung's disease and anorectal malformations.

4. Of all the anomaly groups studied, the period of coping with residual symptoms at home was the most limited for oesophageal atresia patients.

Hirschsprung's disease and anorectal malformations

1. In the remaining surgical conditions, i.e. Hirschsprung's disease and anorectal malformations, residual symptoms were not only very similar, but equally socially embarrassing and difficult for parents to handle at home. In both of these groups numerous examples were found where parents expressed their repugnance for anal dilatations and their disgust at having to work with leaking colostomies. In addition, frequent mention was made of the unrelenting demands posed by day-to-day management of the patient's malodorous toilet-habits. The essence of these findings have not always been adequately illustrated by case material due to the author's reticence in quoting from the very explicit and angry descriptions offered by parents.

Of the 4 anomaly groups studied, the histories of Hirschsprung's disease and anorectal malformation patients clearly reflected that the socially unacceptable nature and the extended duration of residual symptoms had been the most taxing for them. When studying anorectal malformations, other researchers (Dera et al, 1980) have tentatively linked depressive mood in mothers with concern about patient cleanliness, anal dilatations and working with colostomies.

2. Because of the unpleasantness of anal dilatations, many parents allotted to discontinue this aspect of treatment. This was true in both the Hirschsprung's disease and the anorectal malformation group.

3. In the text a number of examples were cited of difficulties experienced by patients in coping with their colostomies in the community. Where the services of stomatherapists were available, these problems were greatly alleviated.

4. In spite of the wide range of barrier creams that were available, skin excoriation remained a problem for most patients. The social implications for the patients are mentioned in both Chapters 6 and 8.

5. In both groups, toilet training became a family issue which affected parents, patients and siblings.

6. Patient adjustment could not be ascribed to surgical results alone. In the text adequate evidence was given that factors such as inter alia patient age, severity of the anomaly, and parent attitudes played an important role in influencing outcome.

7. It became more difficult for parents to discuss symptoms as patients grew older. This was especially true for the majority of cases which were being handled by mothers alone and where the patient was a male.

8. As patients became older, maternal attitudes varied from not really wanting to know how the patient was dealing with his residual symptoms to the other extreme of doing everything for an almost adult patient.

9. In contrast to the parents of anorectal malformation patients, parents in the Hirschsprung's disease group tended to have an oversimplified view of Hirschsprung's disease and the role of corrective surgery. The erroneous expectation was that the removal of affected bowel would result in the complete restoration of healthy function. In the text it was substantiated that the parents did not anticipate the full extent of dealing with the residual symptoms.

B. Community reaction and parent attitude

1. As other researchers (Nevin at al, 1979) have found, it was important for parents' coping abilities to have a positive and open relationship towards the community.

2. As noted by others (MacKeith, 1973), parents were very sensitive to any form of community rebuff, the latter being linked to unpleasant residual symptoms. Where parents continued to reach out to the community in spite of a negative response, the benefits derived were illustrated by examples in the text.

3. Those surgical conditions which had the most socially embarrassing residual symptoms, reflect the largest proportion of closed attitudes. For example, a closed attitude was found in 13% of oesophageal atresias, in 40% of Hirschsprung's disease and in 50% of anorectal malformation patients.

C. Contact with parents of other patients

The most outspoken critics of parent contact were, in fact, those parents who had had direct experience of such contact, notably the cystic fibrosis group. Whereas those parents, namely from the oesophageal atresia group, who had very little direct experience were generally the most favourably inclined to such a source of support.

Although there is extensive support in the literature for parent groups (Boyle et al, 1976; Driscoll and Lubin, 1972; Kerner et al, 1979; Holland and Hattersley, 1980), the present author found that great care should be exercised when introducing parents with similar problems to each other. Some authors have come out in support of the latter point of view (Ineichen, 1974; Field, 1972; Falkman, 1977).

CONCLUSION

While it was difficult to compare cystic fibrosis with the surgically correctable anomalies, the latter showed clearly defined differences between the respective anomaly groups. Proportionately the groups exhibiting the most problems with residual symptoms in the long term adaptational phase, was the anorectal malformation group followed by the Hirschsprung's disease patients with the least problems being found in the oesophageal atresia group.

CHAPTER 7

THE CHILD WITH A CONGENITAL ANOMALY AND SOME FAMILY RELATIONSHIPS

In the present chapter the reciprocal effect between raising the child with a congenital anomaly and family relationships is discussed. Attention is directed at the views of parents on the effects which the difficulties related to the congenital anomaly had on the marital relationship. At the same time this discussion elucidates the role of marriage in helping or hindering the process of coping with residual symptoms in the patient. In conclusion similar aspects of patient and sibling relationships are highlighted.

A. THE MARITAL RELATIONSHIP

A number of researchers have studied the effect of cystic fibrosis on the marital relationship of parents (Chapter 2). Incidence of divorce in a study sample as compared to the national divorce rate is often cited as a criterion in the measurement of such effects. In the present sample six couples obtained a divorce following the patient's birth. This represented one each from the oesophageal atresia and cystic fibrosis groups, two couples from the Hirschsprung's disease and two couples from the anorectal malformation groups. In four instances (representing all anomaly groups), mothers were convinced that the divorce had in some way been related to the patient's difficulties. Although the "divorce rate" in the sample is well below the national mean, such comparisons are not only inaccurate for various statistical considerations, but also not truly reflective of the extent and dynamics of marital disruption in these families.

Parents were asked to evaluate whether their own marital relationship had remained unchanged, had bonded closer or had become more distant due to the patient's difficulties. These results which are depicted in figure 3, show cystic fibrosis parents most often recording negative changes (31%), followed by the anorectal malformation group (20%), the Hirschsprung's disease group (10%) and the oesophageal atresia group (9%). Superficially the results in figure 3 seem to indicate that for all but the cystic fibrosis group, the patients' presence had more often bonded marriages closer rather than having had negative effects. However, close attention to individual responses reveals that strained marital relationships are described in respectively 50% of cystic fibrosis and anorectal malformation histories, 43% of Hirschsprung's disease and 17% of oesophageal atresia histories. In other words, in spite of the strain on marital relationships, some had remained unchanged or had even bonded closer. A brief look at the type of strain described by parents is indicated to further clarify this reasoning.

1. Cystic fibrosis

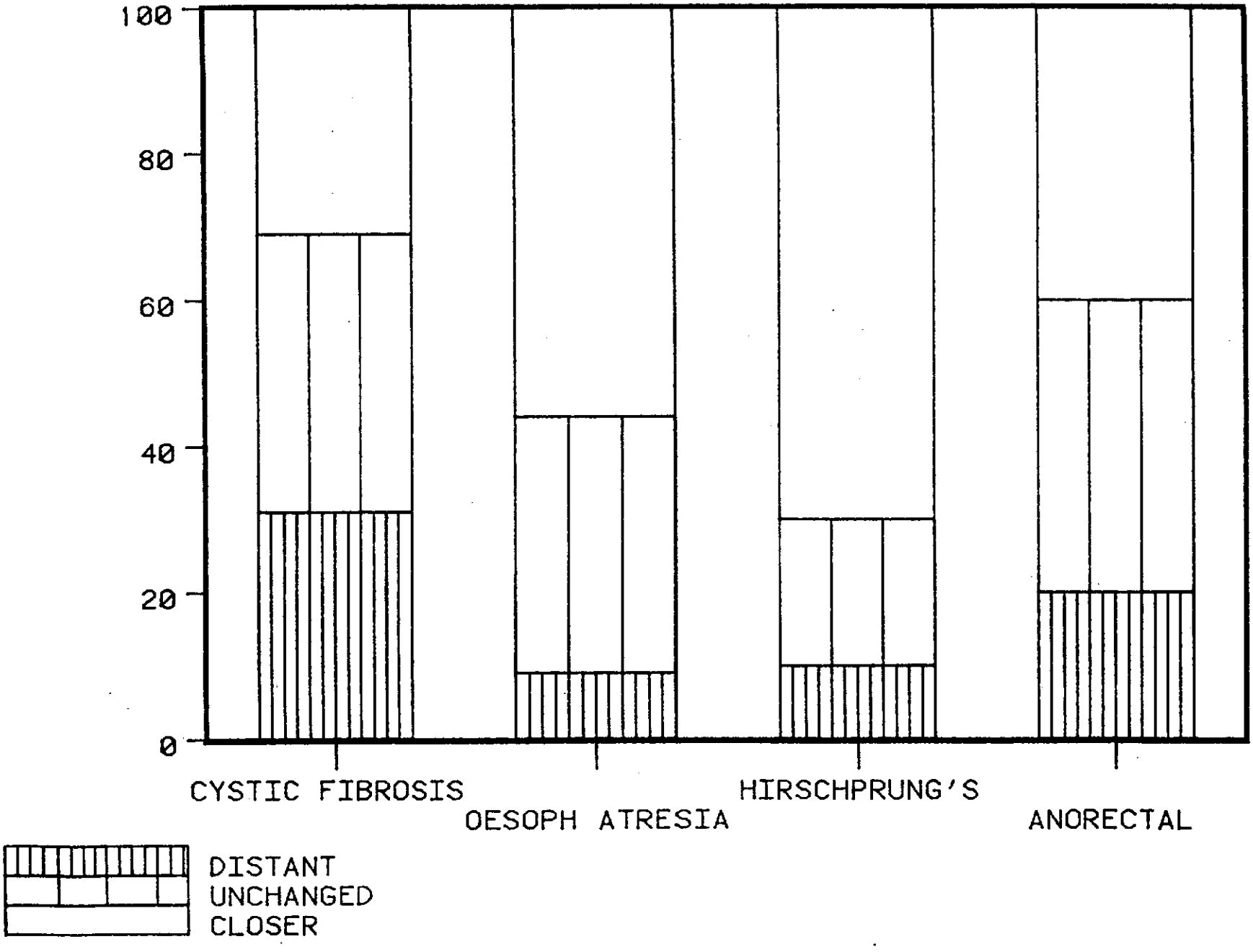
The five parents who rated their marriages as having bonded closer, all mentioned mutual support of spouses in raising a child with cystic fibrosis as the binding factor without explicitly describing strains on the marital relationship.

001 "We stuck together very closely and supported one another."

Three of the six parents with "unchanged" marital relationships, revealed that this status quo had been maintained in spite of severe strain. For example a husband (012) had carried full responsibility for cleaning the house and feeding the children after work at night because his wife had taken to abusing drugs and alcohol while the patient was going through a health crisis. In spite of this pattern repeating

FIGURE 3

MOTHERS RATING OF MARITAL RELATIONSHIP SINCE BIRTH OF THE CHILD WITH A CONGENITAL ANOMALY



itself the marriage was rated as unchanged by both spouses. In another instance stability was maintained only because a father (095) had remained undemanding even though he and the younger sibling had become almost periphereal in the family while the mother devoted full and almost exclusive attention to the patient's well-being. In the third instance (081) disagreements and stormy arguments about the patient's physiotherapy and treatment at home was described as heavily taxing, but not altering the marital relationship.

Five mothers who described marital relationships which had become more distant, referred to one or more of the following contributing factors: the frequent absence of the father (022, 023); his uninvolvement with the patient's illness (022,108); his denial of the severity of the illness (110) and his lack of emotional response to the patient's death (107,108).

The birth of a second affected child was described as the main cause of a dramatic deterioration of their marital relationship in the following history.

> 022
> "With the first one he would help me - sit for hours to get a bit of milk down the stomach, help with the bathing and was closely involved. But with this little child he is restless, anxious, tense. He was never there when I needed him in the sense that I was alone with a sick coughing child for hours on end - then he is away on business - very much away. Dr S said it's his way of escape - hard work. Often we really let fly at one another. I think this illness is finally taking it's toll with us."

The absence of a father was equally felt in a marriage which eventually ended in divorce.

> 023
> "He goes away on business a lot - sometimes for ten days to two weeks in a month. I'm dead serious that he might as well stay away. I really don't need him because when I need him he is not here."

Although physically present, the lack of involvement of a father was seen as detrimental to their marital relationship as the following history reveals.

> 110
> "When a child with cystic fibrosis is born, a marriage either breaks or becomes more cohesive - I don't think there are inbetweens. My own has played see-saw from the one end to the other and I have had to glue it together many a time. Since I have become a Christian I feel stronger to cope on my own - he is always denying that there is anything wrong with B. I wonder whether it is such a blow to his ego to admit to himself that his child has a fatal illness."

Breakdown of sexual relationships during the patient's acute illness and death was described in two histories as taxing to the marriage (107,108). Mothers complained that neither illness nor death had been talked about and that they had remained unaware of their spouses' emotions at this time. With such feelings of separateness they had remained disinclined to continue sexual relationships,(108) "so things are not good between us."

2. Oesophageal atresia

Proportionately, marital relationships in this group showed the least change in the sample both in negative rating (9%) and in reported strain (17%). Marital breakdown in both cases which were rated more distant, had occurred in the acute phase, immediately following establishment of the diagnosis. In the first example (106) mother and child were permanently rejected by the father. In the second instance (083) the already strained relationship (due to the father's alcoholism) received a final blow when mother and baby moved into a separate bedroom. At the time of first interview sixteen years later, the parents were still separated although living in the same house.

The effect for the remainder of this group is well sumarized as follows.

> 018
> "I don't think it really influenced our relationship. Perhaps we are a bit closer, but no dramatic effect."

Other motivations for rating a closer relationship included working together towards the patient's recovery (006, 039, 046, 065), husband's attentiveness during the acute phase of illness (091) and the fact that the effect of marital discord would destroy the patient's chances of survival (052). In essence the attitude in these families can be described as follows:

> 006
> "In that difficult time it was never a case of Dad going out tonight but Mom will have to stay because of P. If Mom and P could not go with him, Dad stayed home as well."

One example was found where lack of emotional support from a father in the acute phase was sorely resented and still remembered many years later (048), although this had not been severe enough to effect any permanent change in their relationship.

In contrast to the other anomaly groups, fathers' involvement with patients had mostly been indirect, i.e. by means of supporting mothers emotionally by their presence at home. Furthermore it seemed that once the marriage had survived the acute phase of oesophageal atresia, the anomaly per se exerted little if any influence on the marital relationship.

3. Hirschsprung's disease

Although half of the marital relationships of the parents of patients with Hirschsprung's disease were rated unchanged, six of these fifteen marriages had only remained so because severe strain had been successfully tolerated. Furthermore, four of the 12 couples who rated a closer marital relationship, equally recorded aspects of Hirschsprung's disease which remained taxing to their marriages.

The reasons given for marital relationships becoming more distant due to Hirschsprung's disease (099,008,004) included the following:

> 099
> "D's illness shook our marriage. Originally because we were both always tired having to get up to him such a lot at night. We didn't have time for each other, we were always too tired to be interested. Every little thing seemed like a big thing. And I still now feel that I have to work harder at it because we still argue about D all the time. I seem to say one thing and he wants to do it another way."

In the second history (008) cited in Chapter 5, the birth of the patient simply hastened divorce in an already shaky marriage due to the father's alcoholism and inability to support his wife. In the last marriage which was rated as becoming more distant, the birth of a second affected child seemed to have stretched a good marital relationship towards becoming negative.

> 004
> "With A's illness we clung together, needed each other because it was a new experience to us - one operation after the other, in and out, having him at home a little while and then back to hospital again. In all this togetherness I think it strengthened us. But with S it was a completely different kettle of fish. The hasles we have today, I still put down to her being born with Hirschsprung's disease. It hit me, but it hit him harder. He lost his confidence in the world. And I needed him just to be his normal self - considerate and sure of his love. But S did not draw us together, in fact it started a very difficult period of our lives."

No clearcut conclusions could be reached about the implications for marital relationships where a second affected baby was born. For example the opposite reaction was found in a history (044) where a once very shaky marital relationship had started drawing closer immediately preceding the birth of a second affected baby. This positive process continued throughout the patient's operations and handling of his residual symptoms at home. In a third example referring to the birth of a second affected baby, the influence on the marital relationship was summarized as follows by the parents:

036
"The second time, apart from some depression, I was never tense or nervous. I think both of us handled it very well. The credit goes to my husband who remained very, very calm. Although we have loved one another since our childhood, this whole thing drew us closer - we became the best of partners in handling Hirschsprung's disease."

In the last example (075,076) of two affected children in a family, parents reported an unchanged marital relationship, but experiencing severe strain. This was also apparent during the combined interview when the father stated that with his help, his wife was coping adequately. She countered by raising his frequent absence from home due to work and sport as causing friction. He reiterated that his own feeling was that he had already sacrificed a great deal of his extra-mural activities to accommodate raising two children with Hirschsprung's disease.

Good marital relationships were often associated with the practical assistance given by fathers (025,034,037,038,059,080,109). Offering understanding when a wife was temporarily unable to continue their sexual relationship (002) or where she was unable to fulfil household tasks (079), were also mentioned as positive factors. In spite of many difficulties it was clear that mutual support, both on a practical and emotional level, played a vital role as a preserving factor in marital relationships. The majority of histories also confirmed that the marital relationship was much needed in order to cope with the demands posed by Hirschsprung's disease.

042
"I must admit having had the support of marriage helped - having had somebody who never said the problem is yours. He bailed me out a lot of times when he saw that I was at my wits end. I don't know how well one would have made out as a single parent."

4. Anorectal malformations

Of the surgically correctable anomalies, the anorectal malformation group reported the highest proportion of both "distant" (20%) as well as strained marriages (50%).

In three (045,066,094) of the four marriages becoming more distant, the core of the difficulty originated or was accentuated in the acute phase of the illness. For example (045), after the patient's birth a father's existing problem of alcoholism became progressively worse. Because the patient had been the only possible male to continue the family name, his wife was assisted both materially and emotionally by her father-in-law. When the latter died, the marriage ended in divorce because the father was unable to support her in any way. In a further example (066) the father's frequent absence in the acute phase originally served the purpose of meeting the additional financial demands of the illness. At the time of interview 3 years later, the mother was still ambivalent about the factors which had contributed towards their unsatisfactory relationship.

> 066
> "Psychologically it drove us apart because I was ratty and horrible. Perhaps it would have been worse if he had not been working so much. I don't know whether it was sheer exhaustion, but because I could not cope everybody irritated me. I did not really want to talk to my husband - even sat in the loo just to be alone. Today I feel even if he was here to help occasionally it would be super. But then he loses his rag so quickly, the child would probably irritate him something terrible and he'll yell at me. When G is sick my husband gets too impatient, I think because he does not know what to do."

The demands posed by the nature and duration of the residual symptoms in anorectal malformations postponed the resumption of normal socializing for most families. One family ascribed marital disruption to the father's insistance on maintaining the status quo in this respect.

094
"He was so irritated, he could not understand my crying and feeling tired. Although I desperately tried to run things normally, I just was not capable of entertaining one more guest. He refused to cut down and that was too much for me. From my side things are probably the same but he ... I don't know what his feelings are."

Only one clearcut example (007) was found where a marrige became gradually more distant although full support had been given by a father during the acute phase of the anomaly. The mother described a progressive indignation of her husband towards the patient's symptoms and personal hygiene. At the same time "he intentionally buries himself in his profession and is totally uninvolved with D."

As in the other anomaly groups, closer marital relationships were often seen as the result of "working on it together" (027,049,090,057). In some instances this implied, at least temporarily, major role changes, especially for fathers.

049
"It disrupted our entire household. My husband suddenly had to do the cooking, cleaning, washing - everything. We felt it was important that I cope with her. It was tiring and stressful but we took it as it came."

5. Some general factors

Some of the aspects mentioned in the individual histories, viz. absence of the father, his lack of involvement with the patient and his support to the mother, need to be given further perspective.

(a) Absence of the father

Absence of the father at some stage of the patient's illness was mentioned as a hardship by 35% of mothers of anorectal malformation patients, 27% Hirschsprung's disease patients, 26% of oesophageal atresia patients and 23% cystic fibrosis patients. The reasons for absence included business, conferences, work courses, border duty and sport. On the other hand, appreciation for the father's direct

contributions towards the patient's well-being was found to be especially mentioned in 22 histories in the sample. Contributions included amongst others forfeiting promotion in order to stay close to the children's hospital, working shifts which would facilitate better home treatment and changing work to obtain medical aid benefits. Such examples were proportionately found in the histories of 41% cystic fibrosis, 22% oesophageal atresia, 20% Hirschsprung's disease and 10% anorectal malformation patients.

(b) Mothers' source of main support

However, when mothers were asked where they had found their main support in dealing with the difficulties of the anomaly, very little correlation with the above is found. Table 30 lists the percentage of mothers finding support from the four sources most frequently mentioned in the sample.

TABLE 30

SOURCE OF MAIN SUPPORT (MOTHER'S VIEW)

Source of support	Cystic fibrosis %	Oesophageal atresia %	Hirschsprung's disease %	Anorectal malformation %	TOTAL %
Fathers	41,1	8,7	43,3	45	34,4
Religion	23,5	34,8	23,3	25	26,7
Grand-parents	11,8	21,7	26,7	10	18,9
Friends	17,6	-	6,7	15	8,9

The foregoing data reveals great ambivalance in the majority of mothers about support of fathers. Although it was much needed, only one-third of the sample singled out the spouse as main source of support. This is confirmed in individual histories where, although a husband was listed as main support, his frequent absence in times of crisis was lamented. Furthermore 23% of mothers in the sample saw themselves as the "stronger person", self reliant or essentially alone when dealing with the difficulties of the anomaly. Where patients became older with unpleasant residual symptoms, mothers occasionally mentioned finding

themselves in a buffer position between father and patient.

(c) Subsequent pregnancies

As an essential part of the marital relationship, the effect of the anomaly on influencing decisions about further pregnancies needs to be briefly noted. From the summarized data (Table 31) it is clear that the least influence was exercised by oesophageal atresia.

TABLE 31

INFLUENCE OF THE ANOMALY ON FURTHER PREGNANCIES

Influence of anomaly	Cystic fibrosis N	%	Oesophageal atresia N	%	Hirschsprung's disease N	%	Anorectal malform. N	%	TOTAL	
Last planned pregnancy	9	52,9	8	34,8	16	53,3	11	55	44	48,9
No effect	2	11,8	14	60,9	14	46,7	8	40	38	42,2
Patient "replaced"	6	35,3	1	4,3	-	-	1	5	8	8,9
TOTAL	17	100,0	23	100,0	30	100,0	20	100	90	100,0

Reaction to the fatal nature of cystic fibrosis is also clearly reflected in the large proportion of explicitly stated pregnancies intended to "replace" the patient. In the similar category for oesophageal atresia and anorectal malformations, both patients had died.

The histories indicated that decisions on no further pregnancies had been taken jointly in the majority of cases, with the fathers being more insistent in this respect. Four examples were found where mothers disagreed strongly enough to use devious means of falling pregnant (005,050) or had stated that they were pining for another baby and were tempted to do so (047,061). The summary of one of these histories (005) depicts a number of considerations when decisions on further pregnancy were made. Mrs Y. for example stated that since L's birth, her husband became the "biggest baby" in the house. His upset

state about L needed more of her attention than anything else. He had made it quite clear that they could not have any more children. L would need all their attention and he feared repetition of Hirschsprung's disease. He insisted on contraception for his wife - something which he personally monitored. Due to their differences about L's illness and her husband's emotional demands, she was of opinion that their relationship was being stressed beyond it's limits. She reasoned that a baby in the family would not only direct attention to someone new, but would also prevent L from remaining the "baby" and claiming excessive mothering. By omission of contraception she fell pregnant. This was confirmed during father's absence while on business overseas. The news was broken during a telephone call from him. On his return home angry feelings had subsided. In retrospect she felt that not only had their marital relationship improved, but that an essential shift away from being so patient-centered had been achieved.

Where no effect of the anomaly on further pregnancies was claimed, some parents nevertheless postponed family expansion, or with cystic fibrosis and Hirschsprung's disease admitted that they had been unaware of the possibility of repetition of the anomaly. Without exception all unaffected subsequent births were regarded as having had a positive effect on marital as well as family relationships.

(d) Financial burden

Financial burden due to the treatment related costs of cystic fibrosis has been associated with strained marital relationships by some researchers (Chapter 2). In the present sample 42% of parents felt that patient treatment and care had never made any difference worth noting to family finances. The results for the respective anomaly groups are

shown in Table 32.

TABLE 32

REPORTED EFFECT OF THE ANOMALY ON FAMILY FINANCES(%)

Reported effect	Cystic fibrosis %	Oesophageal atresia %	Hirschsprung's disease %	Anorectal malform. %	TOTAL %
Some difficulties experienced	53	52	60	65	58
No effects	47	48	40	35	42
TOTAL	100	100	100	100	100

The smallest percentage of parents experiencing some financial strain was found in the oesophageal atresia group where such problems had also been limited to the acute phase of the illness and mostly related to hospital costs. Although a very similar proportion was found in the cystic fibrosis group, their costs were ongoing and not hospital related as treatment was free of charge for these patients. However, with its lengthy pre-diagnostic phase, many families of cystic fibrosis patients had already incurred heavy debts with private consultations before reaching provincial hospitals. Patient diet was expensive and in addition examples were found where no costs were spared to allow the patient to "live to the full" (001, 095). In some instances such privileges were expanded to siblings so as not to cause rivalry.

> 001
> "One of the big problems that cystic fibrosis brought about was financial. It's one of the things that make you the brokest - the maintenance of the disease. I expected A to die at 7 years like I was told. So only a private school was good enough, but then all of them had to go because I did not want them to feel left out".

In the Hirschsprung's disease group 60% of parents experienced some financial difficulties of which half was serious. The latter included examples of a mother taking temporary work with each hospital admission

in order to remain solvent (079) and selling house and business to cover costs, returning to solvency only 6 years later (002). In further examples, exactly half of the family's income went towards paying off debts for a number of years (099) or family expansion was postponed and finally abandoned due to financial strain (087).

Results in the anorectal malformation group where 65% of parents experienced some financial strain, tended to be very similar to those of Hirschsprung's disease. Examples included selling house and moving to lower class areas (050), finding additional employment (066), paying off debt for a period of 18 months after the last operation had been performed (049) and eliciting financial assistance from extended family (070).

Common factors to all anomaly groups were heavy costs of both transport and private consultations where parents lived far afield. Positive effects were recorded where comprehensive medical aid was available. Although it was not possible to establish a direct relationship between financial hardship and marital disruption, it is reasonable to surmise that additional demands were made on marital relationships where family resources had been tapped to such excess.

B. SIBLING RELATIONSHIPS

In the present discussion reaction between siblings and patients will be reviewed in very general terms only, as related by their parents. Intensive research where the effects of variables such as ordinal position, sex, family size and severity of the illness are individually accounted for, fall beyond the scope of this study. The histories confirmed that some siblings remained unconcerned about residual symptoms or the extra attention given to the patient. Yet within the

boundaries of the same family, other siblings reacted to these factors. In summarizing the data, a "problem-orientated" approach was followed, i.e. if any one sibling-patient relationship revealed negative reaction, it was listed as such.

Twenty patients who were an only child to the parents, or had died as a baby, or had had only one sibling under the age of one year, will be excluded from this discussion. The sibling-patient reaction reflected in Table 33 is therefore based on the adjusted frequencies, i.e. the remaining 70 patients in the sample.

TABLE 33

SIBLING-PATIENT REACTION (70 PATIENTS)

Reaction	Cystic fibrosis %	Oesophageal atresia %	Hirschsprung's disease %	Anorectal malform. %	TOTAL %
Concerned and helpful	8	17	14	17	14
No noticeable reaction	15	61	48	33	42
Negative reaction	77	22	38	50	44
TOTAL	100	100	100	100	100

As was expected with the very limited duration of symptoms, oesophageal atresia patient-relationships revealed the smallest proportion of negative reaction (22%). The parents of 77% of cystic fibrosis patients related negative reaction between siblings and patients with declining proportions in the anorectal malformation (50%) and Hirschsprung's disease groups (38%). The nature of difficulties are briefly reviewed.

1. Cystic fibrosis

Attention-seeking behaviour in three siblings, all younger than the patients (102,107,110), was seen as very disrupting to both patients and siblings. In two histories this behaviour continued after the patients had died, but as all siblings had been under 5 years of age, this was regarded as temporary.

Only one example of simulating cystic fibrosis was found in a 9 year old brother of a 14 year old patient (096). He insisted on being given a similar diet while at the same time imitating cystic fibrosis symptoms. Periodically he presented with a number of somatic complaints and refused to attend school as he felt too weak.

Even in older siblings exceptions made for patients were sometimes regarded as favouritism. This was reacted to by either ostracising the patient from friendship groups (108) or quietly going a separate way in order not to cross paths with the patient (095). Other reactions included treating the patient as perfectly normal "but such a bore with all those symptoms" and claiming more attention for their own ailments (001). Many parents expressed concern that siblings had to be subjected to growing up in an environment of illness and family tension due to cystic fibrosis. In one history with two affected children this was described as follows:

> 022
> "She grew up with coughing, sick and even dying children in our home. Her experience of children is suffering. She means such a lot to me, but I think that she worries herself sick about us and her studies are being severely affected."

The above history was also the only example found of concern in a sibling about the possibility of being a carrier of cystic fibrosis.

2. Oesophageal atresia

Apart from one example (040) of attention-seeking behaviour in a 5 year old sibling of a 3 year old patient, uncertainty about possible detrimental affects on siblings, lodged mainly in their mothers. In other words no observable dysfunction had been displayed by siblings themselves, but practical and/or emotional privation was seen to be suffered. In two such instances (065,069) patients were not yet symptom free and still required hospital treatment.

> 065
> "I find the advent of another operation very unsettling - especially when I think of the other children. Last year she had to live with my mother - attend a different school and adjust to a new environment with all its demands for a whole 6 weeks. She had been such a keen girl guide, but just missed her badge because of his treatment. Even in Sunday school she missed her award because of him. One really feels that she is at the bad end of the stick".

In a last example the 19 year old patient (048) had been completely free of symptoms from the age of 2 years. The mother was convinced that her eldest daughter's embroilment with an unacceptable sub-culture was directly related to unfulfilled childhood needs due to the demands of oesophageal atresia. Similarly some currently worrying behavioural characteristics in her 20 year old son were seen to originate from the acute phase when by necessity all attention had centred around the patient, leaving siblings "out in the cold". Only one example of such reported long term effects on siblings directly ascribed to oesophageal atresia was found.

3. Hirschsprung's disease and anorectal malformations

Although proportionately more histories of anorectal malformation patients reported patient sibling reactions than those of Hirschsprung's disease patients, the nature of reactions had been very similar. Verbalized objections to so-called favouritism was found in the under 5

year old siblings of both anorectal malformation patients (060,062,067,088,092), as well as Hirschsprung's disease patients (005,025). Again parents were hopeful that this would be a temporary reaction which would resolve itself as siblings became older and the need for special attention to the patient become less important. However, in both groups an example (002,103) of negative reactions in 18 year old siblings were found, indicating long term reactions in a small proportion of cases. In both histories the parents lamented "neglect" of siblings and together with their expressed guilt feelings, reasoned that siblings reactions were justifiable. The difficulty of handling her own as well as sibling reactions, was described as follows by the mother of an anorectal malformation patient.

> 103
> "At 18 years old she still suggests that B has had more of me. To this day there is a tremendous barrier between them. With such an age gap between them you would think that it would have resovled itself by now. But B used to come between us physically and emotionally. It could have been just a hello kiss to M when I come home from work, but immediately she would have something go wrong to switch my attention to her. Although B now absolutely adores M, M still resents B and the attention she needed those years."

Where patients were between the ages of 8 - 12 years old name-calling by siblings triggered by the presence of residual symptoms, was found to be present in both the Hirschsprung's disease and anorectal malformation groups.

For the sample as a whole, 9 histories revealed that although a very good and helpful relationship was described between patient and older sister, the latter might well have been at risk of bearing responsibilities which were beyond age appropriate level. The presence of such responsibilities were found in the histories of 2 cystic fibrosis, 3 oesophageal atresia, 3 Hirschsprung's disease and one anorectal malformation patient. In 5 of these instances, this older

female sibling was described as the main source of support by the mother in coping with the difficulties related to the patient's anomaly.

The cystic fibrosis histories indicated mainly emotional support - someone to cry with and talk to about cystic fibrosis. The siblings concerned were 13 years and 17 years old and respectively 3 and 4 years older than the patient. In contrast histories of the surgically correctable anomalies showed support by means of direct involvement with the patient in 6 out of the 7 cases and in the last instance indirect involvement by taking over the mother's household duties when the patient required her full attention. In the oesophageal atresia group sibling involvement was of limited duration and restricted to the responsibility of feeding mostly unwilling and slow eaters. In all cases patients and siblings had been under the ages of 5 years and 11 years respectively. The histories of patients with Hirschsprung's disease and anorectal malformations indicated more extended involvement both over time, as well as responsibility in handling the unpleasant duty of cleaning up incontinent patients. Sibling ages ranged from 13 to 19 years.

DISCUSSION

A. Marital relationships

1. The present study confirms the findings of others that complete marital breakdown in the families of affected children was not more prevalent than in the general population (Dorner, 1973; Freeston, 1971; Walker et al, 1971).

2. Out of the six marriages which ended in divorce, four mothers were convinved that the reason for breakdown was in some way

attributable to the anomaly.

3. Although the incidence of divorce was low, a substantial proportion of marriages were strained. This phenomenon was represented in the different anomaly groups as follows:

Cystic fibrosis 50%, anorectal malformations 50%, Hirschsprung's disease 43% and osophageal atreasia 17%. In her study of cystic fibrosis patients, Lonsdale (1978) reported that marital strain was present in 55% of marriages.

4. In all groups the physical and emotional absence of fathers was regarded by mothers as a negative factor. Conversely, a good marital relationship was most often associated with active involvement by fathers in the management of the anomaly. Only one-third of mothers in the present sample mentioned their spouse as the source of main support. This feeling that emotional problems were being faced alone was noted by Esseveld (1973). In this respect Phillipsen (1974) in his study of families with a child with hydrocephalus, reported that a third of marriages could not render adequate mutual support.

5. Between 31% and 40% of the present sample reported that marital relationships became closer as a result of the anomaly. This phenomenon has often been reported elsewhere (Drotar et al, 1975; Martin, 1975; McAndrew 1976 and Kerner et al 1979).

6. The lack of involvement of the father has been noted to have a definite effect on the marriage (Falkman, 1977). Where, however,

fathers were able to forgo traditional male roles in order to accommodate the demands brought about by the anomaly, a closer marital relationship was reported.

7. From the case histories it became clear that the oesophageal atresia groups showed the least dramatic effects on the marital relationship.

8. The lack of communication between spouses about the anomaly was most acute in the cystic fibrosis group. The negative effects of the foregoing on the marital relationship were reported by Kulczycki et al (1969) and Falkman (1977).

9. Where there were further unaffected births, it was regarded as having a positive effect on marital relationships. Where a second affected child was born, reactions were so individual as to defy generalization. These varied from near breakdown to much closer bonding.

10. In the present sample 58% of families admitted to incurring financial difficulties as a result of the anomalies. However, the present study was unable to link the financial difficulties directly to marital disruption.

B. Sibling relationships

1. The siblings of patients with congenital anomalies have often been reported to be at risk of showing behaviour problems and difficulties in coping (McMichael, 1971; Kew, 1975; Lonsdale, 1978; Poznanski, 1973). In the present sample negative reactions in

siblings were most frequently reported in the cystic fibrosis group of patients, followed in declining proportions by the anorectal malformations and Hirschsprung's disease groups. Oesophageal atresia showed the least negative reactions.

2. Any results must be interpreted with caution because reactions which could have been normal sibling rivalry were inadvertently ascribed by mothers to the anomaly. Simeonsson and McHale (1981) have pointed out that accurate assessment of sibling reaction could only be done by direct observations. Furthermore those researchers who have used objective test material found no significant psychological impact on siblings of patients (Gayton et al, 1977).

3. Notwithstanding the above, the concern felt by mothers when interpreting sibling behaviour was nevertheless seen to be very real. For this reason alone these feelings cannot be ignored and in certain instances might even warrant intervention. For instance mothers often verbalized the feeling that siblings had "missed out" - a factor also noted by McAndrew (1976).

4. In 11 cases legitimate parental involvement with the patient's treatment regimes was misinterpreted as favouritism by the siblings. Mikkelsen et al (1978) reported similar findings in their study of cystic fibrosis.

5. Simulating cystic fibrosis symptoms by siblings was noted in only one family. Attention seeking behaviour was seen as a temporary occurrence by parents and was therefore not regarded as serious.

6. A commonly reported problem was the help expected from siblings (Hunt, 1973 and McAndrew, 1976). The present study found 9 examples of instances where the responsibilities carried by siblings were above the age appropriate level.

7. Trevino (1979) drew attention to the risk of depriving "good and helpful" siblings of their normal childhood. In the present study the older sister was found to be especially vulnerable in this respect.

CONCLUSION

The mutual support within the marital relationship was found to be of great importance when coping with the anomalies. The marital strain highlighted can therefore not be ignored in the overall management of the child with a congenital anomaly.

In sibling relationships the present study did not find the same extent and intensity of difficulties as reflected in the literature. More rigorous research design is indicated to isolate those siblings with problems whose source is directly related to the anomaly in order to distinguish the latter from normal sibling rivalry.

CHAPTER 8

LIVING WITH A CONGENITAL ANOMALY: THE SCHOOL EXPERIENCE

The school experience of cystic fibrosis patients as reflected in the literature, was discussed in Chaper 2. A similar handful of studies on patients with surgical conditions were looked at in Chapter 3. From these findings it seems reasonable to accept that no meaningful difference in IQ would be found between the patients in the present sample and a normal school population.

With the exception of oesophageal atresia patients, the majority of the sample in the three remaining anomaly groups were still experiencing residual symptoms when first attending school. From the histories it became clear that school attendance had been the most demanding and injurious phase of coping with the anomaly for the patients themselves. This was the first time when residual symptoms had to be handled by patients on their own outside of the home environment. Unfortunately the uncompromising and inflexible attitude prevalent in the majority of schools, hindered rather than helped the process of assimilation into the wider community.

The present chapter examines the nature of difficulties experienced at school and the way in which they were handled by both patients and their parents. As many of the studies based on test material (see Chapter 2) were very optimistic about school adjustment, the present discussion warrants extensive citation of parental views in order to substantiate our pessimistic conclusions on the realities facing the child with a congenital anomaly at school.

A. CYSTIC FIBROSIS

Of the 17 cystic fibrosis patients, it was possible to study the school experience of only 7 pupils mainly due to the high mortality in this group. As depicted in Table 34, five patients were currently attending school at the time of interview.

TABLE 34

SCHOOL ATTENDANCE OF CYSTIC FIBROSIS PATIENTS

SCHOOL ATTENDANCE	N
Currently attending	5
Attended but died	2
Never attended: alive	5
Never attended: died	5
TOTAL	17

Two pupils had died, one during the course of study and the second ten months prior to the first interview with the parents. Five patients died before having reached school-going age.

1. Handling cystic fibrosis symptoms at school

As in the other anomaly groups, the underlying reason for most difficulties had been the total ignorance of teaching staff of the patient's anomaly. All parents in the sample had paid visits to the school to explain symptoms and treatment to staff, in the majority of cases armed with the brochure from the Department of Health and Welfare on cystic fibrosis. In only one case had this yielded permanent and satisfactory results. In this instance, despite having cystic fibrosis in a very severe degree, a patient had been brought up by her parents as

a perfectly normal child. She took part in tennis and ballet and also played a major role in every school concert. Described as a clever child who enjoyed going to school, she had had the ideal of becoming a medical doctor if she could live that long. When she died at the age of 15 she had completed standard 8, without having missed any one year at school. Because they lived in a little country town and her general practitioner had been aware of her illness, everyone in the community had known the full implications of the disease. She had also known that her illness was fatal, yet had been determined to make the most of her life while she could. In this instance there had been no problem with peer acceptance nor had there been any secrecy at school. This patient had made the most healthy adjustment possible at school, with a full understanding of the severity of her illness.

However, the above history proved to be the exception in the cystic fibrosis group. For example during the primary school years parents had to rely on teachers to administer the pancreatic enzymes at lunchbreak. Although the importance of this medication to digest food and to counteract oily stools had been explained to staff, mothers were again, as found in the pre-diagnostic phase, regarded as over-anxious. As a result medication was either forgotten or wrongly administered. For parents who had vigilantly controlled the treatment regimes at home, such an oversight was unacceptable.

> 001
> "So for years I have kept her reasonably healthy and did not worry too much. But the time that became a worry is when your child has to go to school. When you control everything yourself, it's fine, but the trouble starts when you hand them over to someone else. My child now goes to a private school, because there at least I have some say - I pay and can say what I want. But even there, the cotazyme I gave to Mrs H to give to the child, she put in a glass of milk. You know cotazyme turns milk into vomit and of course she would not take it. So I went in again and explained to someone else how to do it. But I could never really trust anybody there with her - they always forget the cotozyme and then you notice all

> the symptoms are back. Then you realize they don't really bother - this is your problem, you'll have to cope with it."

Teachers equally regarded the regular administering of antibiotics as originating from an overprotective attitude in mothers. Not realizing that children with cystic fibrosis did not acquire any immunity, school staff were careless in these duties and compliance for young patients was poor.

> 001
> "With every new child that came in, she would pick up any kind of illness they might be having. She picked up mouth disease, ulcers, everything, it was very difficult. That is why she has got a very broken up schooling but she has done quite well."

Where patients were old enough to administer their own antibiotics at school, parents often wondered whether they had really been taken, as emotional resistance to treatment existed at home. This resulted in a feeling of losing control of treatment while the patient was at school. In three cases where patients were responsible for taking their own antibiotics, the only alternative for parents had been to avoid high risk situations of exposure to infections.

> 110
> "R is an excellent soccer player and is always included in the team as he is a good goalmaker. When it rains, however, I refuse to let him play in matches which are away from home. Once they played at X, 100 kilometres from here. They were totally drenched in rain and sat in the bus wet to the skin. Soon after that he was down with another chest infection".

In the above history even repeated explanations of the nature of cystic fibrosis symptoms and the required precautions failed to elicit appropriate reaction from staff. Although the parent's ruling had been that the patient not participate in away matches if it rained, his sportsmaster vowed that he would "cure the sissy".

> 110
> "He was due for another match, when the rain started coming down in torrents. I went to fetch him, but he refused to get into the car wanting to play the match. Eventually when we did go home, it

> ended with his tossing and turning all night, disturbing everyone. I finally got up and went to his room. He confided that he was afraid of the hiding which awaited him at school the next day. He would really have preferred coping with a chest infection rather than the hiding."

The finding of Burton (1975) that children with cystic fibrosis fear both physical violence and verbal abuse, could explain the patient's preference to rather tolerate a health risk situation than not to conform. Yet even in situations where extra-mural activities had not been enforced, patients preferred to participate in order to gain peer acceptance.

> 096
> "I feel if she wants to do anything, she can do it but the only thing I am not for is swimming. When they go to the beach in summertime, it is fine. She will get dried and dressed and that is a different matter but not at school. They have got cement floors in the changing rooms and her hair gets wet and stays wet in the classroom. She invariably gets a chest infection and I can assure you it happens over and over again. On the next day the child is sick. But then she sort of felt odd because the other children were all in the water. But I said to her 'what would you prefer, would you prefer to be sick or would you prefer to swim?'

All cystic fibrosis children in the sample were keen school attenders. The majority resented having to miss days at school to stay home when they were not well. This seemed to stem partly from the desperate striving to be regarded as totally normal. Most parents endorsed these efforts but then only to the point where the patient's health would not be affected.

> 110
> "It worries me a lot that R is trying so desperately to keep up with his friends at school. He wants to attend every camp and outing they have. But with all the junk foods going on these camps he comes back very ill. But when he is there he refuses to watch his diet and eats everything the others eat. The whole family pays for this when he gets back."

Ambivalence between guarding the patient's health and jeopardizing the intensive efforts at conforming to the peer group, was described as follows by the mother of an 11-year-old patient who had just gone

through an acute phase of illness for which she had been hospitalized.

022

> "This week Wednesday an educational camp starts for all the standard three's. All of them usually attend for two nights, one night in a hut and one in a farmer's barn. Now I ask you, she was discharged from hospital last week, she is merely skin and bones. The one thing that has kept her going was looking forward to this camp. How can I say no to her, but how can I say yes? Must I close my eyes to the very likely chance of another chest infection, or shall I give her a straight 'no' ? What must I do?"

The nature of this ambivalence is best understood when it is borne in mind that school attendance was intermittently interrupted for most cystic fibrosis patients during bouts of illness at home or in periods of hospitalization. These absences not only influenced academic achievement, but also hindered socialisation and thwarted the patient's attempts to gain peer acceptance. During their absence new friendships were formed and contact with the "in-group" was lost, as the following illustrates.

> 022
> "Ann was her very best friend who knew her so well. When she felt ill and had pain, Ann would rub her back for her. It was almost pathetic to see the way she cared for her. There are only 9 girls in a class the rest are boys. However, since the last part of her illness the bond with the group seems to have weakened and her friendship with Ann who has found another best friend, has petered out".

In two instances (001,110) parents had changed schools until a satisfactory settlement had been reached, even though this involved considerable upheavel for their families.

> 110
> "Because of his cystic fibrosis, R is now attending his third school. We lived in X village but the damp there was so bad for him, we had to move. I was sorry because he had coped reasonably well in the school there. The second school was very small - only a handful of pupils and only 5 boys in his own class. One child in this group did not like him with his skinny appearance and bouts of coughing. Life was made so unpleasant for him, that we decided to move again so that he could attend a bigger school. Now I think he has adjusted quite well for the last two years.

Three examples were found where the discussion on cystic fibrosis

between parents and teachers resulted in the patient being confronted by his peers with the fact that he was going to die one day. In one history it was described as follows:

> 026
> "It happened three years ago. P's teachers told his classmates not to fight with him, as he would die any day. Naturally, his best friend told him the following day. That is when his bedwetting started, which is still a problem today. I just left it at that - the harm was done but the next year when he changed teachers, I explained cystic fibrosis to her again and referred to what had happened the previous year. I do this every year, with cystic fibrosis brochures, the lot and since then things have been going quite well for him. But that incident was a major first set-back. That child knows that he is going to die".

In a further example an 11-year-old patient (022) was mocked about "going to die anyway" by a pupil who had overheard two members of the P.T.A. discussing cystic fibrosis, following the mother's visit to the school. Although at school the patient responded bravely, she broke down at home and sought the mother's reassurance that she was not going to die of cystic fibrosis. At school, staff members had remained unaware of the incident as the parents had preferred not to pursue the matter.

In the last example (001) the mother, after an equally negative experience, recommended that schools never be informed that cystic fibrosis is fatal, as too many complications could arise which patients would not be emotionally equipped to deal with satisfactorily.

2. Educational status of patients with cystic fibrosis

Superficially very little difference could be seen between cystic fibrosis patients and healthy pupils. As Table 35 indicates, six of the seven patients experienced no serious academic problems: all had been average to above average in class work and five had been active

participants in school sports at the time of first interview.

TABLE 35

EDUCATIONAL STATUS OF CYSTIC FIBROSIS PATIENTS: PARENTS VIEW

Factors evaluated	Yes	No	TOTAL
Academic problems	1	6	7
Sport participation	5	2	7
Interfering symptoms	7	0	7
Special school attended	1	6	7

Two patients had already been in a too advanced stage of the illness to participate in sport. More than half (4) of the parents felt that their child was not reaching full potential, even when bearing in mind the limiting nature of cystic fibrosis. The histories revealed a very heavy investment from both patients and parents in order to maintain a standard of coping which would be regarded as normal.

B. OESOPHAGEAL ATRESIA

Of the 23 patients with oesophageal atresia, 11 had not yet attended school as they were too young and two had died before reaching school-going age (Table 36). Ten families reported on the school experience of patients, eight of whom were currently attending school at the time of study. Two patients had left school, one to marry and be a housewife. The second patient enrolled at a technical college for a post matric course.

TABLE 36

SCHOOL ATTENDANCE OF OESOPHAGEAL ATRESIA PATIENTS

SCHOOL ATTENDANCE	N
Currently attending	8
Left/completed school	2
Never attended: alive	11
Never attended: died	2
TOTAL	23

1. Handling residual symptoms of oesophageal atresia at school

For the majority of patients with oesophageal atresia, postoperative symptoms had resolved before formal schooling commenced (see Table 25, Chapter 6). Three patients had the occasional problem of ridding themselves of a food bolus in public, when it developed. However, these patients managed well, without undue anxiety, and in some instances (064) reacted by jesting about occasional choking in public. From the histories it became clear that socialization problems could not in any way be related to residual symptoms of oesophageal atresia.

2. Educational status of patients treated for oesophageal atresia

Academic difficulties were reported for three patients. In two instances (016,106) similar problems were experienced with healthy siblings. In the first history the patient was married with one healthy offspring. Her mother related that she had always been slow at school, very nervous at examination time and had been relieved to leave school after completing standard 6. None of the three healthy siblings completed their school education.

The second history revealing academic problems (106) was investigated by a local welfare organization. Their report described a family which was frequently on the move, leaving a string of debt behind them. With the concurrent frequent change of schools, the 14-year old patient and her brother had fallen behind progressively in their school work. Both children twice repeated school standards. When last interviewed, the patient had been enrolled at a special residential school in order to counteract the unstable family background and her learning problems. The only residual symptom of oesophageal atresia was her very slow eating.

Academic problems in the third instance (064) were to a large extent pushed to the background by this patient's excellent sport achievements. At the time of first interview he had been chosen as super sportsman of the year for his school. Other achievements included being chosen for the provincial trials of both swimming and soccer and playing school team tennis and cricket. Besides playing an active role in school concerts, extra art classes were arranged for him as he had excelled in this area as well. In spite of his academic difficulties, the impression was gained that his parents were satisfied that he was leading a well-rounded life.

TABLE 37

EDUCATIONAL STATUS OF OESOPHAGEAL ATRESIA PATIENTS: PARENTS VIEW

FACTORS EVALUATED	YES	NO	TOTAL
Academic problems	3	7	10
Sport participation	9	1	10
Interfering symptoms	3	7	10
Special school attended	1	9	10

From the foregoing discussion it is clear that the small proportion of learning difficulties as reflected in Table 37 could not be attributed to any hindering influence of residual symptoms of oesophageal atresia. Sport participation was reported for 9 out of 10 patients and no socialization difficulties related to oesophageal atresia were found.

C. HIRSCHSPRUNG'S DISEASE

At the time of study, 18 of the 30 patients with Hirschsprung's disease were attending school (Table 38). Two patients were working after having gained a matriculation certificate and a third patient had left school, unable to cope. The results of school experiences of 21 patients with Hirschsprung's disease are therefore discussed.

TABLE 38

SCHOOL ATTENDANCE OF HIRSCHSPRUNG'S DISEASE PATIENTS

School attendance	N
Currently attending	18
Left/completed school	3
Never attended: alive	8
Never attended: died	1
TOTAL	30

1. Handling residual symptoms of Hirschsprung's disease at school

The parents of five patients (24%) had throughout the school career been satisfied with the overall progress and social adjustment which their children had made at school. The common factor in all of these histories was the fact that no symptoms had been revealed to the peer group or other pupils at school. This was possible either because patients had been virtually symptom-free, or together with their parents

had made every effort to conceal the anomaly. This group included four patients with short and one with long segment Hirschsprung's disease.

In contrast to cystic fibrosis, the residual symptoms in Hirschsprung's disease proved to be more embarrassing and inhibiting because of the socially unacceptable nature thereof. Sixteen patients (i.e. 76% of those attending school) had been subjected to unpleasant incidents at school. These problems centred around the embarrassment caused by staining, smelliness, flatulence, borborygmi or diarrhoea. These patients and their parents experienced periods of extreme difficulty in coping with the school experience. As will become clear from the histories, some parents remained unrelenting in their efforts to support and steer the patient through a school career until they could finally regard adjustment as satisfactory. Other parents preferred to do what they could at home, and although they had some suspicion that difficulties with symptoms were being experienced at school, refrained from active intervention. A common factor to both the attitudes mentioned above, was the fear of parents that the patients would be unable to cope at school. The attempts to accommodate unresolved symptoms were described as follows by the father of a patient.

> 082
> "A is 7 now and I dread the day when he'll have to go to boarding school. We have sold the one farm to move nearer to town so that my wife could drive him to school and back. Children, especially the smaller ones can be bitterly cruel - and I know he is going to suffer. When I think of the embarrassment - that offensive odour is enough to make the heads turn."

While parents were doing much at home to try and curb symptoms during school hours, the feeling was that little flexibility could be expected from school routine in order to accommodate symptoms.

042
"My biggest anxiety was school and what would happen there. Schools are ridiculous, they have got certain times that they are only allowed to go to the toilet. Thus far, touch wood, he has managed to cope pretty well. He still has to go to the toilet every night. We wake him up and now it has become habit a few times at night."

060
"He is 11 now and does well at school - he had 85% aggregate for standard 3. He plays his rugby and participates fully at school. My part is to get him up before 6 am every morning and to get him to empty his bowel. This works most of the time, although he has had the occasional accident at school. Yet I don't want to interfere at school because one hopes that it will improve."

In some instances parents tried to use unhappy incidents at school to motivate patients to control bowel habits. Whereas some histories reflected a positive response, others revealed the opposite as the following exerpts respectively illustrate.

080
"C had one very bad incident in primary school. Unfortunately he passed faeces in his pants in class and everyone saw this. He was terribly ridiculed on the playground because the story went like wildfire. He refused to go back to school. I spoke to the headmaster who explained to the whole school at assembly exactly why this had happened and the teasing stopped. This changed him. Previously he tried to hold back when a call to go to the toilet came. Now he responds immediately to his own warnings and manages to stay relatively clean. I am almost sure he had no further accidents in secondary school."

100
"J is now 15 and has become very resistent to my insistence about emptying his colon before going to school. The only defence I have is to remind him of the day when he had also lapsed and this had ended with him soiling his school desk and chair. It was such a mess that the whole class knew about this. The true effect of this incident I will never know because he never talks about it. Well, the whole class knows now, but whether he is still ridiculed, I don't know. Nevertheless, he has stopped all extra-mural activities. He loves reading and seems to be quite content with his own company."

Many parents remained unaware of how patients were managing at school even though evidence of some incontinence was found. As in the previous excerpt, some signs of adjustment difficulties also became apparent in

the following example.

> 109
> "In his first year at school he had a couple of accidents here and there. He used to come home about every day with his underpants smeared. Gradually that seemed to die altogether. Now at 8 years his underclothes are clean. In this time he never complained about children teasing him, but he started stuttering. And they do really tease him about the stuttering - terribly."

For the patients themselves colostomies were difficult to hide and handle effectively. As the following histories illustrate, parents' approach to the problem differed widely. In the first example the young patient was literally left to fight his own battles.

> 074
> "D is seven but still in sub A. They said he was more in hospital than at school and failed him that year. This year they said he is doing well, but his bag is still a problem. You see, the doctor said we must bring him up like a normal child and let him play sport. But accidents happen. Like the other day things started running down his legs when the bag was leaking. The children started mocking because they did not know what was wrong. He came away crying. They grab hold of the bag and say ' what is that?' But now he fights with the bag on. You should see the big guys he takes on. Whereas they used to hurt him he now tells us 'don't worry, tomorrow I'll knock them up!'"

In contrast to the foregoing example, other parents actively intervened in order to ensure a successful school career. For instance a patient's (005) good academic results as well as his sports achievements were seen as the outcome of planning hospital admissions to coincide with school holidays. Added to this, regular feed-back to school staff was maintained viz. showing the new colostomy to the headmaster with an explanation of its function and the need for free access to toilet facilities in order to prevent unpleasant accidents. Yet even such efforts did not prevent the same patient from being punished for not wearing the correct gear for physical training. On this occasion he had intentionally remained in his school shirt in an attempt to hide from the rest of the class a colostomy which was not quite clean. Whenever a

new teacher arrived or the patient had been moved to a different class, more visits to the school became necessary, as missing a swimming class due to diarrhoea or frequently leaving the room, resulted in punishment on a number of occasions.

Whereas the abovementioned attempts were handled with resigned humour, other parents responded to such incidents by moving the patient (038) to a private school where symptoms were accommodated with greater flexibility. In this instance the patient had managed to complete matriculation without having further embarrassing accidents.

In 9 of the 16 cases where incidents did occur, mothers visited schools with explanations of Hirschsprung's disease. Yet the impression was gained in all cases that this would have been avoided if possible.

> 035
> "I try not to go to school with his Hirschsprung's business, because I think symptoms don't worry him that much, except in summer with the fruit season when frequency becomes a problem. At the beginning of each year I motivate him to talk to his class teacher to be given permission to leave class when necessary. If after 2-3 weeks he has not managed to do so I invite her around and do it for him."

Only one example (037) was found where a patient had left school after having failed standard 2. Although she had been placed in a special class due to a low IQ, inability to handle unpleasant symptoms and the peer group ostracism, ended her limited school career.

2. Educational status of patients with Hirschsprung's disease

For the Hirschsprung's disease group as a whole, six patients (29%) had experienced academic problems (Table 39), three of whom on one occasion

failed to pass their standards. During psychometric testing, only one (037) was found to be of low intelligence.

TABLE 39

EDUCATIONAL STATUS OF HIRSCHSPRUNG'S PATIENTS: PARENTS' VIEW

Factors evaluated	Yes	No	Total
Academic problems	6	15	21
Sport participation	11	10	21
Interfering symptoms	16	5	21
Special school attended	1	20	21

At the time of interview, 11 patients (52%) were actively involved in at least one type of sport. A further 6 patients had attempted playing sport, but eventually given up, mainly due to fear of close contact with others in the face of symptoms and the lack of privacy when changing into sportsgear.

As was the case with the academic aspects of school, parents again invested heavily in time and effort to ensure continued participation in extra-mural activities. This is well illustrated by the history of a patient attending a private school.

> 038
> "He tried swimming, but his bottom hurt so much, it put him off completely. Although he played hockey and did boxing, he could never really go on school outings involving overnight stays. In standard 10 the housemaster came to me and said we must try, it is his last chance. So I went with them on the bus, and wherever we stayed he and I stayed together. We stayed with private families all the way and it was always 2 boys to a family. Well, instead of 2 boys, it was always him and me. During the day, he sat with his pals on the bus and doing his own thing and nobody was any the wiser. I stayed in the background and did not interfere and allowed him just to get on with it.So at least he had something like that, it was so super for him".

Other patients insisted on doing as much as possible on their own, not without the risk of running into difficulties. For instance where a patient (005) had successfully been on a week-end rugby tour, for the first time managing without any parental assistance. On the way back, his colostomy came free in the bus. At the first stop he got rid of the bag without anyone noticing. However, he had no more replacement bags. To counteract the odour he sat himself in the back of the bus and opened all the windows. Only his closest friends realized his predicament and together they managed to pass off the incident as a joke.

From the foregoing discussion it is clear that many extra demands were made on Hirschsprung's disease patients in coping with the school experience. Very little understanding was shown on the part of school staff, even where contact had been made by the parents. In such cases little continuity was found in accommodating the patient with his symptoms.

D. ANORECTAL MALFORMATIONS

As shown in Table 40, ten patients in the anorectal malformation group were either too young or had died before having reached school-going age.

TABLE 40

SCHOOL ATTENDANCE OF ANORECTAL MALFORMATION PATIENTS

School attendance	N
Currently attending	10
Never attended: alive	9
Never attended: died	1
TOTAL	20

Ten patients were attending school at the time of study, with only two reported as having experienced learning difficulties.

1. Handling residual symptoms of anorectal malformations at school

In the first instance (088) the seven-year-old patient of low IQ was enrolled at a special school. He was of poor social background where the family struggled with problems of unemployment, debt, alcoholism and ill-health of the mother. Amongst the main problems, poor scholastic achievement of this patient was not of great concern to his parents. His admission to a special residential school was accepted with great relief. In the second history revealing learning problems (090), the year in which the patient failed, was attributed to hospitalization. Subsequent difficulties were ascribed to residual symptoms.

> 090
> "She tries to join everything at school. Lately some stories that she has a 'funny vagina' have done the rounds at school, resulting in staring at her and whispering about it. This upset her tremendously - at 12, 13 years this is too bad for her."

As was found in the Hirschsprung's disease group, a large proportion of patients (7 out of 10) experienced painful incidents at school due to their symptoms. In five cases parents visited the schools with

explanations of the nature of the anomaly in an effort to counteract ridicule or ostracism of the patient. In two cases where such difficulties were faced by a seven and eleven year old respectively, the incidents seemed to have been of less serious nature. Nevertheless, the impact on the patients had been sufficient to modify their behaviour connected to the symptoms as the following illustrates.

> 024
> "It is only his peculiar way of walking when his buttocks has a rash, then the boys ask him why he had such a funny protruding buttocks. This really upset him and then he started walking on the outside of his soles which looked even more ridiculous. But presently he never even has an accident at school any more. Whether this is connected to the times that he has his meals or, whether it is some miracle, I dont know. Surprisingly enough from 7 in the morning till 3 in the afternoon, he is beautifully clean, there are no problems. But the minute he gets home, the time for having accidents is there. He relaxes too much and realizes too late that he has to run for the toilet. At school I think it is his pride that keeps him from having an accident, even prevents him from asking the teacher to be let out of class."

In the second example, a seven-year-old managed to keep himself acceptably clean by regularly changing a wad of cottonwool in his underpants. In addition an extra set of underclothes was taken to school to ensure that no odour would give away his problem. His meticulous efforts ended abruptly when other pupils started peering under the toilet door to see what he was doing there so frequently and for so long. On noticing the relapse in his personal hygiene, his mother decided to intervene. An explanatory letter from the surgeon was followed by a visit of the mother to the school. At the time of study this action seemed to have achieved the desired effect: a three month period of no accidents or symptoms at school had passed. The patient since then had been able to restrict his personal hygiene routine to those times when other pupils were not in the wash rooms, because he was then permitted free access to the toilet during class hours.

Especially where fathers had not been closely involved in the handling of symptoms, boys did not discuss difficulties experienced at school. In some cases problems only became apparent when it had also developed into an emotional issue for the patient. For instance a patient (007) who had gone through primary school without ever referring to the intense emotional trauma caused by having 'accidents', only revealed his fears a year before completing his school career. His father had in the past always responded with anger towards any signs of residual symptoms. Because his father had withdrawn more and more into his successful private practice, his son was left without any paternal support.

Although his mother had remained sympathetic, she found it increasingly difficult to raise such personal matters with him. Just prior to a whole day sports gathering at school, he broke down and confessed to his mother that he could not face the day. Although she had suspected a few difficulties, her impression had been that it was reasonably tolerated. The magnitude of the problem for him had come as a total surprise to her.

The case histories show that patients felt more at home in the academic aspects of school where little close socializing was required than in extra-mural activities. These were the times when symptoms became most apparent and difficult to handle. The reticence to discuss difficulties at school and the added stress of extra-mural activities are well illustrated by the history of a 12 year old in standard 4.

> 027
> "This is a great worry for him I think more than we have realized. He tried to hide it, of course, but it's evident in the fact that he had not got friends. It's very sad but he has'nt. He is a difficult little child. The other kids have children coming here

> to sleep, close buddies sort of thing. He can't do that. He'll come to us and say, 'I've got rugby today, what am I going to do?' And then we realized it's been bothering him the whole year. He does not want to play rugby at school because he is expected to shower with all the other boys afterwards and he is not the same as the other boys. He might just even have a stain on his pants. So he has this constant worry at the back of his mind all the time. Then, as soon as we knew about it, my husband said he'll speak to the teacher involved and sort this out. But it's the same thing with his cricket. They expect him to shower afterwards. So we now discovered he does not participate in sport, he stopped playing rugby and cricket and it is so important for him to become involved. My husband's talking at school did not help a lot. You see all the boys know about him because there is the odd occasion he might smell a bit and children are very cruel. The other day he said, 'Mom, when I'm in high school, I hope my problem will be sorted out because then I can start afresh ...' You see, that says a lot. In his way he wants to start anew. All the boys he feels, know that he is like this, they dont want to have anything to do with him - kids are funny. He might have a stigma attached to him. As he is growing up he is becoming more sensitive about it. It's the stage now when he so desperately wants to be like his buddy. He is trying to get himself involved in other things, thank goodness. We have encouraged it. He copes at school, he's quite bright and that we don't have a worry about. He's got a good head but I think that it is not enough. If you are not one of the top ten cricketers or rugby players, you are inclined to be pushed into the background today."

The excellent and supportive family background of the patient in the abovementioned example would probably ensure that his school career be completed in spite of existing difficulties. However, an example to the contrary was found in the history of an 18-year-old (045). Although this patient had never referred to any incidents at primary school, his mother recalled his angry outbursts about why he had to have this anomaly, unlike other pupils. In secondary school he had a very difficult school career, sometimes refusing to attend and at others coming back an hour later because he did not have the courage to face the pupils who were making fun of him. She then realized that he was being ostracized due to bad odour. This she blamed on teachers who would not excuse him frequently enough during early classes to prevent smearing. If this were allowed he would be able to go free of accidents for a whole school day. He persevered till the second term of standard

9 when he gave up and enrolled himself at the permanent force. He discussed his difficulty with his officer-in-command, requesting only one exception to be made for him: permission to be allowed frequent use of the toilet until 9 am. With this being granted, his personal hygiene changed overnight.

Although superficially some patients were coping well, parents were very aware of the extensive efforts made on their part to maintain a normal and well adjusted front.

> 094
> "He is a bit shy, even towards me and will not talk about this openly. He really is very quiet about this. I think he realizes that this is his problem and that he'll have to live with it. One could not really say that he takes this with humour but he has a good life according to his age. He is a litte rascal and has got an infectious laugh yet he can be so aside and lonesome sometimes. The animals are really his friends. He does not make friends easily and when one watches him when he mixes, I always realize that he talks a little too loud, laughs a little too loud and superficially one would think that he communicates well. He tries very hard on the sports field as well, runs in the group, usually loses, plays average tennis but where one really notices is the fact that his principal told me that he has a much higher IQ than his brother has. But his marks in school are so much lower."

2. The educational status of patients with anorectal malformations

The educational status of anorectal malformation patients is summarized in Table 41.

TABLE 41

EDUCATIONAL STATUS OF ANORECTAL MALFORMATION PATIENTS: PARENTS VIEW

Factors evaluated	Yes	No	Total
Academic problems	2	8	10
Sport participation	5	5	10
Interfering symptoms	7	3	10
Special school attended	1	9	10

In spite of the difficulties discussed, 80% of patients with anorectal malformations had managed to obtain academic results which were satisfactory and 50% to participate in at least one type of sport. Many patients had interfering symptoms, yet the major hardship emanated from the unyielding school system which offered very little support to the efforts of patients to effect and maintain normal adjustment.

DISCUSSION

1. With the exception of oesophageal atresia patients, the majority of patients in the three remaining anomaly groups were still experiencing residual symptoms when attending school.

2. The uncompromising and inflexible attitude in the majority of schools was found to be the single most damaging factor which hindered the patients' attempts at normal school adjustment.

3. In all the anomaly groups, the ignorance of teachers about these conditions and their residual symptoms gave rise to this unyielding attitude.

Cystic fibrosis

1. Although cystic fibrosis patients in our sample were found to be keen school attenders, school authorities could not be relied upon to administer vitally important medication.

2. A further problem peculiar to the cystic fibrosis group was the

loss of peer contact as a result of intermittent periods of absence from school.

3. As found in the literature (Stephan and Biener, 1978) cystic fibrosis patients in the present study too, were pushed to their limits both on the sportsfields and during physical training.

4. The eventual death of the child with cystic fibrosis was inappropriately handled at school.

Oesophageal atresia

1. Where patients with oesophageal atresia did experience learning difficulties, this was in no way associated with residual symptoms.

Hirschsprung's disease and anorectal malformations

1. In contrast to cystic fibrosis, the residual symptoms in Hirschsprung's disease and anorectal malformations proved to be more embarrassing and inhibiting because of their social unacceptable nature. Seventy-four percent had been subjected to unpleasant incidents at school caused by staining, smelliness, flatulence, borborygmi or diarrhoea.

2. Parents' attempts to facilitate normal schooling ranged from finding a private school which would accommodate residual symptoms, to active intervention at the present school, to simply ignoring the problem, i.e. hoping that the child would cope on his own.

3. Active intervention by parents at school, i.e. explaining the anomaly, providing literature or by demonstrating the patient's colostomy, yielded only temporary results. No parent could effect a permanent arrangement to accommodate residual symptoms, which could have, in most cases, been controlled by simply permitting the patient to visit the toilet freely.

4. Peer ridicule and ostracism, described by researchers of other anomalies (Dorner, 1976) were also found in the present sample. This could also have been minimized by a simple measure, i.e. full access to the toilet, as described in the previous paragraph.

5. Patients were found to be especially vulnerable due to the lack of privacy both in the locker room and on the sportsfield. Although patients attempted to participate in extramural activities, many efforts were futile because of these problems. This in turn, resulted in a one-sided school career consisting only of the academic component.

CONCLUSION

Findings clearly show that irrespective of their anomalies, patients were able to cope amongst a normal school population. However, a number of factors within the inflexible school system prevented these patients from realizing their full potential both in the academic field and in extramural activities.

CHAPTER 9

THE LOSS OF A CHILD WITH A CONGENITAL ANOMALY

In the previous chapters the difficulties experienced by families since the birth of a baby with a congenital anomaly were discussed. In this overview, which followed a more or less chronological pattern, the special demands posed during the different phases of adjustment to the anomalies were highlighted from birth through school attendance of the patient. However the discussion of psycho-social implications for these families would be incomplete without closer scrutiny of the small proportion of families where patients had died before reaching adulthood.

In the present chapter the special needs of the 11 families where patients had died are briefly discussed as found in their case histories. Because the numbers are small, generalizations are hardly possible. However findings serve the purpose of indicating both the positive and negative aspects of losing a child with a congenital anomaly.

A. CHARACTERISTICS OF PATIENTS WHO DIED

The 11 patients who had died were representative of all the anomaly groups studied (Schedule 7). The four patients with surgically correctable conditions all had associated severe multiple anomalies and died within the first week of life, with only one having had some surgical intervention. The seven patients with cystic fibrosis died between the ages of 6 months and 15 years.

SCHEDULE 7

PATIENTS WHO DIED: DIAGNOSIS, AGE AND SEX

DIAGNOSIS	RESEARCH NO	AGE	SEX
Cystic fibrosis	095	15,9 yrs	F
	108	15,0 yrs	M
	102	5,4 yrs	M
	022b	3,5 yrs	M
	071	1,8 yrs	F
	107	0,8 yrs	M
	096b	0,5 yrs	F
Oesophageal atresia	014	7 days	M
	077	2 days	M
Hirschsprung's disease	004	2 days	M
Anorectal malformation	013	4 days	M

Due to the very short contact between parents and patients in the surgically correctable group, it was anticipated that the emotional trauma of their loss would be limited both in duration and intensity. However, individual histories revealed that much emotional hardship had been experienced in both groups. In the present discussion some similarities and differences will be highlighted.

B. THE DECISION OF WHETHER OR NOT TO TREAT

Parental involvement in the decision whether or not to vigorously intervene with life-saving or life-prolonging treatment for the dying patient, was the most distinct emotional trauma in 2 of the 4 surgical correctable conditions. A vast literature exists on the moral and ethical issues of selection for treatment and whether active

intervention could be justified in certain cases (See Chapter I). Such aspects are beyond the scope of the present discussion which will simply concentrate on the reported effects on parents.

In two instances where Down Syndrome, together with oesophageal atresia and anorectal malformations respectively, were diagnosed, the decision whether or not to actively treat was discussed with fathers shortly after birth. In both instances the histories revealed that in retrospect fathers were intellectually secure that they had made the correct decision. This was related 15 months later, as follows, by one father:

> 013
> "We realized what the whole situation was all about. He informed us about the test results. We knew the extent of .... Dr. D had given us a pretty good idea as to what it would entail in our future with operations, the clinics etc. He was going to come around that evening to discuss the whole matter with both of us. But prior to him coming, and before anybody had said anything, I had decided personally that it just was not on at all. When Dr. D. came round that evening, I said to him, 'Before you go any further, we have already decided that we are not prepared to operate or at least we are not prepared to give consent for operation'."

A similar intellectual reaction was recorded from the second father 4 years after the patient's death.

> 077
> "In spite of everything, I never doubt that I have done the right thing."

Both fathers motivated their decision in similar vein: the burden of raising such an extensively handicapped child would be too much for their wives. In addition, social mobility played a major role in the first case (013).

013
"My wife is at the peak of her career, and I felt that the child would have been an incredible psychological burden - it would have been too much. Apart from being a professional person myself, I have in the past had all kinds of restrictions in my life - my folks got divorced, my mother was an alcoholic trying to bring up two children on her own. Yet my brother and I made our way up in life - God knows how - till we reached a good level in life, which we really battled to get to. I personally landed up marrying a very good woman - somebody well educated, somebody who is able to conduct herself in pretty any sort of environment. Also on a social basis we have done very well for ourselves through a lot of pure hard work. I just felt to have gone right back because of the child, was not on. I thought about what would happen and there is no doubt about it: we would have had to pull back. It would have changed our lives completely and totally. It would have made undone all we have worked for."

In the second example the presence of mental retardation added to the unacceptable nature of an alternative decision being made about not treating the patient.

077
"My wife could not have carried the burden of a child with so many defects. We as a family could not have coped. Both my son and daughter are of above normal intelligence and I cannot see any place for a child who is mentally retarded and with other defects in our family. From that point of view I have made the right decision, but ...".

Whereas parents in the two foregoing examples had broadly followed the same lines of reasoning on an intellectual level, some differences in their emotional reactions were found. During the first interview, which a father (077) requested to be with him alone, he confessed to still feeling guilty, four years after the patient's death.

077
"I take it you know what passive euthanasia is? Well, I was party to that and therefore to the death of my child! ... I have made the right decision but from a religious point of view, what I have done can never, never be justified."

During this interview the father became tearful and emotionally extremely upset. He explained that the decision not to treat had been taken by him without consulting his wife, who had given birth by Caesarean section and could therefore not have visited the hospital. When the patient died 35 hours after birth, he still had not told her about "my part in the decision-making". He decided to withhold this information from her altogether as he was not sure how she would handle this. At the time of our first individual interview he expressed a fear that his wife would suspect something if a research interview was done with her and he was still uncertain what her reaction to this information might be. He acknowledged that he was not only protecting his wife from further emotional hardship but at the same time feared that such knowledge would alienate her from him - she might not condone the action he had taken at the time.

In the second instance (013) 15 months after the patient's death, the father felt emotionally secure that the correct decision had been taken.

> 013
> "My wife fell pregnant 5 months after T died and I feel this is a very healthy situation."

From a religious standpoint this father felt no compunction and at the time of interview indicated that as a family they were "beginning to look ahead". In contrast to the previous example, his wife had been involved in the decision-making process and all knowledge had been shared. He nevertheless admitted that he had played the major role and therefore took responsibility for the action taken.

013
"I must emphasize that my wife was under great mental strain at that stage. It hit me later on that throughout that early period I was making the decisions and she was following. But with me there was never a question, never a question that the right decision had been taken".

In both examples the time between deciding for no active intervention and awaiting the patient's death proved emotionally draining, even in retrospect. Both fathers decided that further viewing of the infant would only contribute to the emotional hardship of their wives. In retrospect one doubted the correctness of this decision.

013
"There were always mental pictures of the poor little chap lying in the hospital, waiting to die as it were. We looked at it in that light - it was a human reaction, I suppose. It was a very bad period, but nothing was said between the two of us then - only much later she would say that she had been straining to just slip away on the quiet and go and see him. And I was the same, in spite of our resolve not to go. Afterwards - 2-3 months later - it was much worse."

077
"I will always bear the responsibility of my child dying of hunger and thirst without the medical means of saving his life, because I decided that they should not treat any further."

From the interview it became clear that some misconceptions were present. Furthermore, this father's feelings about the patient's death had not resolved.

077
"People see me come and go and probably think the matter is settled, but in one's heart the child remains irreplaceable in spite of the many defects he had."

As will be shown later in the discussion, this family had not completed mourning four years after the patient's death, as the father greatly

inhibited any overt signs of sadness in his wife and other children. Although the researcher had initially been refused an interview with his wife, he expressed a feeling of great relief following the discussion and requested a combined interview at home on condition that "the euthanasia not be discussed."

In the examples cited above, both fathers stressed the fact that every opportunity had been given to them by surgical staff to discuss problems or raise questions. At hospital the matter had been well handled and the decision taken had been their own. Yet after both interviews much relief was expressed at discussing the patient's death, and in retrospect gaining more insight as well as confirming the correctness of action.

In the third example the patient died before the decision of whether or not to treat had been taken by the parents. This was a patient with oesophageal atresia and multiple anomalies, who in addition was found to have an extensive heart condition which precluded surgical intervention at that stage. However, a cardiac surgeon had tentatively discussed the alternatives of whether or not to operate. A year later the parents feelings were summarized as follows.

> 014
> "It is a decision that has to be made by doctors with experience in these type of cases. What would happen if we had said no, don't operate, and then later find that children with similar conditions have survived and eventually got better? They shouldn't have told us this. I still thought to myself 'how can you ask me to give consent to killing my baby?' I mean probably if you are in the medical field it's different but you know for us it was a hard thing, it was our own child."

(In the examples cited above (013, 077, 014), parents had been of social classes II, III and IV respectively.)

For the parents of cystic fibrosis patients who died, termination of treatment never really became a central issue. Only two examples, (071, 107) were found where parents in the terminal phase of illness felt that continued treatment had merely prolonged the dying process unnecessarily. In both cases they had not verbalized this at hospital, as staff had been so bent on treatment, that "our attitude would have looked so unthankful and uncaring." Apart from the resentment expressed by parents where they were informed that the child had already died, other histories generally revealed satisfaction with terminal care at the hospital.

C. COPING WITH THE DEATH OF THE PATIENT

1. Preparing for the loss of the child

Mothers of children with cystic fibrosis often mentioned in passing that they had tried to prepare themselves for the loss of their child.

> 102
> "In the last two years I have been thinking what will it be like when it finally happens. I suppose I have been trying to prepare myself for that day all along. I always thought I would just go mad. But it was not like that - I think I was calm."

In one instance the emotional as well as practical preparation for the patient's death was described as follows:

> 096
> "I took her to hospital and they admitted her straight away. When I came home, I washed the cot out and folded it up. Together with the pram. I put it in the spare room. When my husband came home he asked, 'What did you do with A's cot and pram?' I said to him, 'I know A won't come home'. He said I was talking nonsense, the

> doctors were busy with her, but I knew. I consoled myself then already, so that when the knock came after that, when the knock on the door came, I was prepared for it."

In retrospect, some parents realized how irrational the "preparation" in their minds had been. For example (107) anticipating the news that the patient had died when the parents had heard a car door closing. Yet it was quite clear that the news would be conveyed by telephone. In a further example (002b) a mother who had already lost a child with cystic fibrosis described her feelings with the death of the first, and preparing for losing her second child.

> 022b
> "Gradually it penetrated that I was going to lose him, he was weak and emaciated. The thought of losing him made me aggressive and I would not accept it. I started fighting for him with everything humanly possible."

Having lost the first child, she describes an intellectual awareness that the second child would also die of cystic fibrosis. Yet acceptance in advance was thwarted by the difficult way in which children with cystic fibrosis died.

> 022b
> "My reason tells me that cystic fibrosis will take her - so what are my fears? My anxiety about her coming death is the suffering, because no child with cystic fibrosis softly closes the eyes and goes. It is a cruel hellish suffering when they die. Otherwise I could have accepted it - she has to go some day, so, eyes softly closing, gone - take her Lord! But with cystic fibrosis it's not that easy, I dread the day."

Only two examples were found where death nevertheless came as unexpected to parents. All other parents (5) of cystic fibrosis children who died, anticipated the imminence of death as hospitalizations became more frequent and infections more resistent to treatment. One mother (107)

described an unbearable sadness with the realization that she would not be taking her child home again - she knew that this would be the last admission and cried at his bedside.

> 107
> "The sister pulled me up short with 'you're not the only one with a sick child around here! Pull yourself together or go home - you are upsetting the other parents'. That day I dried up completely and was just left tearless and hollow when he died a few days later."

The history of a relatively healthy cystic fibrosis patient who was still alive at the close of the study, revealed that the mother had forced herself to watch the process of deteriorating health and dying of another patient in an attempt to prepare herself for this coming ordeal.

> 026
> "I got to know C when she was in hospital. I used to visit her every week and phoned her every day, right up to the end. It was something terrible to see the suffering of that child and how she finally died. It was horrible. I really could not stand to see her deteriorate day by day. All the time I was looking at her I saw my own child, but something compelled me to go through with that. I just had to do it, because I said to myself: 'This is waiting for you, you will also have to go through this one day'. I will not recommend that anyone ever do it. No mother must ever see the terrible way in which a cystic fibrosis child dies."

Of the cystic fibrosis group as a whole, 10 histories revealed the depressing effect on other parents when news got round that a patient known to them had died. In contrast to the history above, seeing or hearing about the death of patients was avoided and not regarded as preparation for the death of their own child.

> 110
> "On the ward when I asked about certain patients I was always told that they have passed away. I just could not face that, it left me in the doldrums."

001
"It is too depressing how the patients die one after the other. It is something that I try and avoid."

2. Parents presence with the dying child

The majority of parents (6) had been present when their child died. In five instances the parents had only been informed by the hospital after the child had died. In three instances this had happened by omission of hospital staff and in 2 cases by choice of fathers that the dying infant not be viewed. In retrospect all mothers regretted their absence and described feeling inconsolable at the loss of the last opportunity to be of help and comfort to their child.

096
"I felt very uneasy at home and wondered whether A was alright, because I felt all upset about her. But they said if anything was wrong they would let me know. Afterwards Dr M said 'Look, you could have done absolutely nothing for her because she was too far gone already.' So I said I understood. I just felt sad because I wanted to go and be with her. It's only natural to feel that way."

107
"The saddest was that after all the suffering he had to die alone. As his mother I should have helped him in that most difficult time. I grieve having lost the opportunity for doing just that."

In one instance (071) social work intervention at the last minute had secured the privilege of being with the dying child. The mother had sought assistance with the plea, "If she is going to die anyway, please just let me into the I.C.U. in time!" Attempts at resuscitation had been so intense, that the parents who were waiting outside had almost been forgotten. Their presence, for less than a minute before the infant died, was related in retrospect as a source of consolation to themselves.

Four examples were found where fathers had been opposed to being present when the patient died. In two cases where they had nevertheless been reluctantly present, much relief was expressed in retrospect that they had "gone through with it".

102
"It was strange to see my husband there because he didn't want to be. I knew I had to be. On the Monday night he just walked in - it was 11 o'clock and he had just had a feeling and decided to come. So he was also there and today he is still relieved that he had come."

In one instance (108) the patient himself had given up hope that his father would come. By aggressive intervention of the doctor he was fetched just in time to say good-bye to his 16 year old son who died a few minutes later. Once more relief was expressed in retrospect that he had been able to be a "good father" to the end.

3. The meaning of the patient's death

Finding a reason why their child had to die became relevant for parents following the patient's death. The majority found some meaning in a religious sense. These reactions varied from a continued blind and unquestioning faith to becoming somewhat sceptical, as the two examples respectively illustrate.

096
"The night before she died our minister came and said to me, 'M, are you satisfied whatever the Lord declares?' So I said yes, and he prayed. I have accepted her death and that she was only loaned to us for that short while. There are lots of people who can't have children. We were blessed with children and if they are taken away you must just accept it."

004
"For a long time I felt almost turned away from religion. I have never offered a mass for him yet, like most Catholics do."

Other parents had found some meaning during a funeral service, while others referred to the benefits derived from visits and counselling by a minister of religion. It was clear that the death of a child had often taxed and shaken a previously firm faith.

> 102
> "I am not a regular church goer, but when C was so ill I phoned the minister and asked him to pray God to take him quickly. And then he died that afternoon. When the minister came over I told him that I doubted and I have never doubted before that there was a God. I asked why did it have to happen to us? You get baby bashers, murderers and nothing happens to their children. So he said that God chooses two people, a whole family or even whole community to get a child like that. And then he takes it away, not as punishment, but to teach everybody something. When he said this, it just seemed so true in our case."

> 014
> "I think what helped the most was the service when he was buried. In that sermon it answered most of the questions in my mind. It was hard for all the family to accept his death. The priest explained that he was loaned to us for a short while and I think it helped us to start accepting his death."

Total negation of religion as an avenue for finding meaning for the patient's death was found in one instance. This history (071) revealed a mother's fear of becoming embroiled with the "religious fanaticism" of her husband's family. She insisted on a private cremation attended only by her husband and herself before informing relatives of the patient's death. The parents alone scattered the ashes on the ocean before returning to their home a few hundred kilometres from the hospital.

Although most parents had described that "there was no sudden rush to church or religion" (013), it was clear that for the majority religion

had offered much by way of emotional strength and consolation following the death of their child.

4. Replacing the dead child

In two cases the deceased child had been the last born to the family, with both mothers having been sterilized. Five examples were found where pregnancies were planned, immediately following the death of the patient. Sometimes mothers directly verbalized the fact that this was done to fill the void of losing the patient, as the following excerpt indicates.

> 096b
> "It was about 3 weeks after that when I fell pregnant. I did not want to wait too long to have another baby because I wanted to occupy myself with something. She also had so much baby clothes that was never used before she died. Now when I look at B's face I can still see S, the way she looked in her little coffin - the two faces are one."

Seeing a keen likeness in the child born to "replace" the patient, was found in other histories. In one instance the mother had seriously considered naming the baby after the deceased patient.

> 014
> "He was born exactly 11 months after M's death and has completely taken his place with the grandmother. I really wanted to call him M as well, but my husband said it would not be fair on the child. But he looks exactly like M and everything in him reminds me of M. As he progresses, I always wonder how M would have done it."

The mother in the example cited above, added that she was always tempted to say that the second born was her first child, in order to forget the loss they had suffered. In another history, a baby born 4 months prior to the patient's death served as "replacement" and brought some

amelioration to the loss, although the pregnancy had not intentionally been planned as such.

> 022b
> "When S died H was 4 months old, - a boy, identical, they could have been twins. Then I said, God is good to me. In other words, although the feeling of loss was there, I found peace of mind and acceptance when I realized that I had a healthy son. And they were as similar as two drops of water. Exactly the same being, similar eyes, hair, everything - only he was bigger. Two months after his death I could feel myself recovering: I had a son exactly like the one I had lost."

Replacing the dead child nevertheless took its toll by way of periods of serious doubts and depression during the nine months of pregnancy. Although mothers had been well aware of the genetic risks involved, they had chosen to "take a chance of a healthy child". In one instance (071) the "replacement child" was also affected. In a further two instances (022b, 096b) more pregnancies followed with two more children affected with cystic fibrosis being born.

As can be expected, where a baby of different sex to the patient was born, such detailed comparisons were not drawn by parents. All histories revealed that from parents' point of view the child born after the patient, was regarded as a consolation and as easing the process of mourning the patient's death.

5. Coping with grief

The majority of parents reported going through a phase of heightened activity in an attempt to counteract the feeling of loss. For example some mothers reverted to spring-cleaning the home after terminating char

services (108) or working during the day and reading at night (071). Others joined sports, gymnastics or physical fitness classes (077, 107, 108) or simply socialized excessively in order not to be alone and depressed (004). Depression and loss of confidence were expressed in three cases (095, 004, 107).

Whereas all the parents of cystic fibrosis patients had welcomed the interest of friends together with the opportunity this offered of talking about the deceased, the reverse was found to be true in 3 of the 4 families of patients with surgical conditions. In the latter cases interest was regarded as inquisitiveness and therefore resented. These parents were sometimes tempted to lie in order to prevent further questions (013, 014). The impression was gained that discussion was acutely avoided in those cases where a decision against active intervention had been taken by parents. In one instance (077) where the father had taken this decision without his wife's knowledge and he himself still had unresolved religious conflicts in this respect, discussion was curbed even in the immediate home environment. The history revealed intense strain in the mother, who only cried and mourned her child when she was alone at home in order not to upset him. Four years after the neonate had died, emotional upset of the spouses compelled the researcher to abandon the research schedule in favour of a therapeutic interview. In spite of this the father had not yet felt secure enough to share with his wife the decision he had taken and his religious conflicts surrounding the baby's death. Following an individual interview with the father, much relief was nevertheless expressed for having had the opportunity of sharing his load with someone - "A load I have carried alone for four years."

Without exception, all the families where patients had died, expressed appreciation that "someone from the hospital" had taken the trouble to visit them. In one instance (014) where the visit was paid less than 3 months after the patient had died, some surprise was verbalized that it had taken so long. Furthermore these families had been more than willing to co-operate with research. It nevertheless became clear that following the death of a child in hospital, parents had felt the sudden severing of all contact with the hospital as the loss of an avenue of support. At the same time the histories confirmed that many areas for rendering emotional support and answering questions existed which could ease initial coping with the loss of the child. For example in three instances (071, 096b, 014) where post mortems had been done, further questions arose in parents' minds which they felt reticent to raise after contact with the hospital had been broken. It was verbalized as follows at the close of the first research interview at home:

> 014
> "We expected that someone would contact us just to tell us finally whether something we had done could have caused his condition. You know after they cut him open they would perhaps discover more. Anyway, now we must just go and spread his little ashes, then everything is done."

The above history revealed the parents' feeling of having gone through all the necessary steps to ascertain that they had not in any way contributed to the patient's death, after which mourning rituals could be completed. The fact that the post mortem could serve to relieve the parents' mind of vague feelings of worry was mentioned again (096b) where the mother felt that her confidence had been somewhat restored by the diagnosis being thus confirmed.

096b
"They asked me if they could do a post mortem and I thought, well, why not? If they can save someone else's baby, it should be done. Now I also have more confidence and I feel more people should consent to them being done."

No overt examples of incomplete mourning viz. maintaining the patient's room as a shrine (Kerner, 1975) were found in the sample. However one example of inhibited mourning in family members was discussed earlier.

D. DISCUSSION

The decision to treat the patient

The study would be incomplete if the special needs of the 11 families where patients died were not documented. However, because the numbers are so limited, extensive generalization is precluded.

1. In reaching the decision whether or not to actively intervene with lifesaving or life-prolonging treatment, fathers had, in retrospect, been intellectually secure that the right decision had been made. Their primary consideration had been for the well-being of their wives, i.e. the emotional and practical burden of raising a severely handicapped child.

2. Other factors influencing their decision were the unacceptable nature of mental retardation and the disadvantages of handicap to upward social mobility.

3. From the vast volume of literature on decision making where critically ill infants are concerned, two schools of thought have

developed - one which places the responsibility in the hands of a hospital based committee, the other, in the hands of the parents and physician (Watchko, 1983). The present study indicates that parents alone did not feel equipped to make a decision.

4. It was clear that, following such a decision, emotional support was sorely needed. The period of greatest vulnerability was while awaiting the patient's death. At this time further decisions as to whether or not to visit, view or hold the baby, had to be taken. All three families involved in this decision making distanced themselves from the patient and elected not to visit. The ambivalence evidenced in the text is well described in the literature (Limerick and Downham, 1978; Walker et al, 1971).

5. In the one instance where both the mother and father had been involved in the process of decision making, communication and mutual support of spouses were facilitated and improved. The opposite was found where a decision was made by a father alone. In the latter example the mourning process was greatly inhibited, the spouses remaining silent, not wanting to upset one another (Ross, 1978).

6. The present study revealed that religious doubts and some misconceptions surrounding the death of the patient culminated in guilt feelings in the parents.

Preparing for the loss of the child

1. The individual manner in which families prepared for the loss of a child is described in the text.

2. In certain cases similarities with anticipatory mourning as described in the literature was found (Fulton and Gottesman, 1980; McCollum and Schwartz, 1972; Naylor, 1982; Friedman et al, 1963).

The presence of the parents at the dying child

1. In the present sample 6 parents had been present when their child died and 5 had not. All parents agreed that being present was important for both patients and mothers. The literature confirms this importance in order to facilitate mourning (Bourne and Lewis, 1984).

The meaning of the patient's death

1. Religion played a definite role in influencing the understanding of parents of the death of their child. Reactions varied from the very positive to the very sceptical.

Replacing the dead child

1. Detailed examples were found where mothers hoped that by falling pregnant they might fill the void of having lost the patient. Other authors have pointed out the detrimental effects to healthy mourning, of hurrying into a further pregnancy (Bourne and Lewis, 1984).

Coping with grief

1. Various forms of heightened motor activity following the death

of the patient was found in the present study. This phenomenon is in keeping with Lindemann's (1944) classic study of bereavement.

2. Following the death of a cystic fibrosis patient, parents evidenced an overwhelming need to talk. This stands in stark contrast to their earlier reticence to discuss the approaching death of the patient.

3. Where a decision against active intervention had been made, parents actively avoided talking about the patient's death to friends and acquaintances. A possible explanation for this reticence might lie in unresolved feelings surrounding the decision to allow the patient to die.

4. In the majority of parents whose children had died, there was an expressed need for continued contact with the hospital as an avenue of emotional support. This was especially relevant where postmortems had been done and many unanswered questions remained in the minds of parents. This finding is in keeping with the conclusions of others (Berger, 1978; Valdés-Dapena, 1979).

CONCLUSION

The discussion has pointed towards the need for the overall management of the child born with a congenital anomaly to extend beyond the last admission to hospital. Areas were highlighted where families could benefit by further contact with the hospital following the loss of a child.

CHAPTER 10

MAJOR FINDINGS AND RECOMMENDATIONS

As set out in Chapter 1 the primary aim of the study was to gain insight into the immediate and long term psychosocial difficulties, if any, of families where a child with a life threatening birth defect was born. A comparison was made between those patients requiring surgical intervention, i.e. oesophageal atresia, Hirschsprung's disease and anorectal malformations and those suffering from an hereditary condition, i.e. cystic fibrosis.

HYPOTHETICAL BASES OF THE STUDY

It was firstly hypothesized that a smaller proportion of the families of patients with life threatening surgical conditions would have benefited from the services of social workers than the families of cystic fibrosis patients. This hypothesis was upheld in that social work services were most often rendered to families of cystic fibrosis patients (25%), whereas the families of oesophageal atresia patients were proportionately least often helped (4%). The families of patients with Hirschsprung's disease and anorectal malformations respectively, availed themselves of social work services in 10% of cases.

The nature of social work intervention had been mainly material assistance and largely limited to lower middle class families. However, in Chapters 6 to 9 it was adequately illustrated that a much larger proportion of families had been subject to a far broader spectrum of psychosocial difficulties which could have benefitted from social work intervention. The recommendations will highlight some of the areas requiring such intervention.

The second hypothesis was that a core of difficulties experienced would be common to all four of the diagnostic categories studied, while other difficulties would be peculiar to the specific type of anomaly and to the families involved.

This hypothesis was substantiated in the findings summarized in Chapters 4 through 9. The areas of high risk and the specific problems pertaining to each anomaly group will be further highlighted by way of the recommendations made to alleviate these difficulties.

The third hypothesis anticipated that the duration of psychosocial difficulties related to the anomaly would be most limited for the families of those patients where an anomaly was corrected, leaving the minimum of residual symptoms. Oesophageal atresia was the anomaly group most fully corrected leaving the minimum of residual symptoms. The psychosocial problems of this group were largely limited to a period of intense anxiety in the acute stage of the anomaly. As was shown in Chapter 8, by school-going age these patients exhibited no overt psychosocial difficulties pertaining to oesophageal atresia, which was in sharp contrast with the many psychosocial difficulties still present in a sizeable proportion of the other anomaly groups when attending school (Chaper 8). This was particularly true of those patients where residual symptoms had still been present. The details supporting this hypothesis are outlined in Chapter 6.

FINDINGS AND RECOMMENDATIONS

As set out in the aims of this study, the present Chapter will make recommendations for enhancing the coping strategies of patients and their families, with the view to social work intervention specifically

geared to the identified needs. As the study has revealed needs which fall beyond the scope of social work intervention, some general recommendations equally affecting the well-being of the families will be made.

In view of the specialized nature of recommendations made about the role of the social worker, it is furthermore imperative that social workers dealing with families have a thorough working knowledge of the specific anomalies and the nature of their demands. This implies keeping abreast with both medical and social work literature pertaining to these anomalies and forming an integral part of the team handling these patients. Ideally the social worker should form the liaison link between patients, their families and the medical team.

I. PRELIMINARY SYMPTOMS AND LEARNING THE DIAGNOSIS

The most important factor underlying the problems experienced by the parents immediately following the birth of patients in the study, was the delay in diagnosing the anomaly. Although it is anticipated that currently fewer patients will experience a lengthy prediagnostic phase, the main thrust against these problems remains to obtain a rapid diagnosis. While this is first priority, the other measures serve merely to alleviate the circumstances created by this failure to establish a rapid diagnosis.

A. IN THE MATERNITY HOME

1 Where staff were suspicious of a birth defect in a baby, symptoms were handled by them without informing the mother of these

difficulties. This resulted in the unclarified absence of the baby at feeding times which in turn aroused suspicions in the mother, sometimes of gross deformities. Parental speculation was often so distorted and uninformed that the emotional drawbacks of not viewing the infant, outweighed any possible advantages of withholding information from them. A further problem experienced by the mothers at this time was extreme loneliness at being separated from their babies.

It is therefore recommended that

(i) no feeding times be omitted without explaining to the mother why her baby is not brought to her;

(ii) information be shared with parents even where uncertainty still exists about the exact nature of the defect;

(iii) social work services be offered with the view to practical and emotional support during this time of loneliness.

2. Whereas staff suspicion was followed by treating symptoms, mothers' suspicions were not taken seriously and could not elicit any constructive attention. Much hardship could have been prevented and a rapid diagnosis established if mothers' suspicions had been supported by the necessary investigations.

It is therefore recommended that

until proven otherwise, mothers' suspicions be taken as valid indicators of ill-health in their babies.

3. (a) Although preliminary symptoms might have been treated in the maternity home, a large proportion of babies were nevertheless discharged without a diagnosis having been made. This problem was compounded by the failure to make an adequate referral for

further investigations after discharge.

(b) As far as the parents from the lower socio-economic group was concerned, an avenue of return to the maternity home was made available in case preliminary symptoms should recur after discharge. The study revealed that this measure aided successful diagnosis within a reasonable period of time.

It is therefore recommended that

even where preliminary symptoms responded to treatment in the maternity home, further recourse back to the maternity home be made available to parents.

4. The study has shown that where suspicion of ill-health was followed by direct referral to the Children's Hospital, the prediagnostic phase had been most effectively shortened.

In view of the above it is recommended that

where possible staff at the institution of birth who are already familiar with the baby's symptoms, should make a referral to a centre equipped to establish a diagnosis.

5. The dearth of communication from staff to parents with respect to suspicion of ill-health within the patient, resulted in prolonged separation of mother and infant when directly transferred to a hospital for confirmation of a diagnosis. This separation gave rise to the following problems i.e.

(a) mothers not viewing their babies at all where the birth had been by caesarian section;

(b) irreplaceable loss where the patient had died before the mother was discharged from the maternity home;

(c) difficulty in easily establishing a mother/child relationship;

(d) lasting hostility towards the medical and nursing profession.

It is therefore recommended that

(i) viewing of the baby should be allowed irrespective of the extent and severity of the anomaly in order to:

- counteract fears of gross anomalies
- ameliorate feelings of loss if the baby should die
- facilitate the development of the mother/child relationship;

(ii) any hostility towards the medical profession be explored during social work intervention in order to better equip parents to deal with the subsequent demands of hospitalisation and treatment.

6. (a) For mothers, the direct transfer of the baby to a hospital resulted in a further period of traumatic separation which, in turn, lead to many of them discharging themselves prematurely from the maternity home.

(b) Fathers were often involved in transporting the baby to the hospital. Furthermore they were burdened with the responsibility of keeping the mothers informed about treatment decisions and progress of the baby.

(c) In a few instances marital relationships were such that fathers had been unaware that the child had been born with a birth defect.

In view of the above it is recommended that

(i) the separation be ameliorated by establishing regular contact between the hospital and the mother in the maternity home - a duty which can be shared between the social worker and sister of the neonatal ward;

(ii) regular instamatic photographs of the baby be taken in order to

- ameliorate separation
- facilitate development of the mother/child relationship
- counter-act phantasies of gross anomalies;

(iii) social work support be extended to the fathers.

B. IN THE COMMUNITY

Establishing a diagnosis in the community proved to be a lengthy and demoralising experience for the majority of parents. Once again parental concern about the patients' symptoms were dismissed too easily and without the necessary investigations. Instead, symptoms were interpreted as being the results of inter alia poor training, overprotectiveness, or to be of psychological origin. The failure of medical practitioners and specialists to refer to a centre equipped to make a diagnosis when they themselves were unable to do so, contributed to this delay. The contrary was found to be true for parents in the lower socio-economic class. In these instances a successful diagnosis was more readily forthcoming because of direct referral to the Children's Hospital.

A number of problems could be directly ascribed to this delay in diagnosis.

(a) Parental confidence was seriously eroded during the course of seeking a diagnosis in the community.

(b) Because symptoms were sometimes attributed to faulty training or psychological factors, the mother/child relationship was jeopardized.

(c) Failure to make a rapid diagnosis evoked intense agression in parents against doctors with the added loss of confidence in the medical profession in general.

In view of the above it is recommended that social work intervention following diagnosis of the patient should take note of the above factors. In subsequent interviews these areas should be consciously explored with a view to

(i) working towards restoring parental selfconfidence;

(ii) arranging for maximum opportunity for handling the infant when eventually hospitalized with active support in establishing a healthy mother/child relationship;

(iii) exploring parents' feelings directed at the medical profession and providing opportunity for ventilation and catharsis in order to establish a constructive working relationship between doctors and parents;

(iv) interpreting parental difficulties to medical and hospital staff;

(v) the social worker taking an active mediating role until the doctor/parents relationship has been established.

C. HOW PARENTS WERE INFORMED ABOUT THE DIAGNOSIS

1. The study revealed both inter and intra-diagnostic differences in parental response to being told about the patient's birth defect. Parents of cystic fibrosis patients having the greatest proportion of delayed diagnoses, also most often expressed relief at being given a diagnosis. This temporary reaction was found to a lesser extent in the Hirschsprung's disease and anorectal malformation groups. An acute fear of losing the patient was a common response of parents in the three groups of surgically correctable anomalies. This response was most frequently and acutely felt in the oesophageal atresia group. In contrast the parents of children with cystic fibrosis found it extremely difficult to verbalize fear of the patient's death.

   It is therefore recommended that following diagnosis social work intervention should

   (i) deal with the abovementioned common responses i.e. fear of the patient's death;

   (ii) at the same time differentiate treatment goals in order to deal with the problems which are peculiar to the specific anomaly groups.

2. The majority of parents in the sample had been satisfied with the way in which they had been informed about the diagnosis, notably so in the surgically correctable anomaly groups. In all groups the attitude of the "teller" was found to be of

primary importance, overriding most of the identified negative factors.

Although the task of breaking the news to parents has traditionally been assigned to the diagnosing surgeon or paediatrician, indications were that more flexibility needs to be practised in this respect.

It is therefore recommended that

(i) the diagnosis be given with the social worker being present in order to provide for both the factual and emotional issues;

(ii) where conflict between specific parents and informant might exist, the news be broken by a different member of the hospital team in the presence of the diagnosing doctor;

(iii) the parents again be seen the following day after being informed of the diagnosis to answer any questions as parents often do not grasp the facts at the first session.

3. The study indicated that the fatal nature of cystic fibrosis was accepted with difficulty and that a small proportion of parents then regarded the "teller" as being blunt. At the same time the booklet on cystic fibrosis was not well received by the majority of parents.

(i) The foregoing confirms the recommendation that the social worker be present when parents are told the diagnosis.

(ii) It is further recommended that the social worker extend emotional support to the parents on a continuous basis, whilst at the same time reinforcing factual information by repetition where regarded as necessary.

(iii) In view of the negative parental response it is recommended that the cystic fibrosis brochure be initially withheld and handed only selectively to parents requiring printed information. Handing out the brochure should never be seen as a substitute for direct, personal communication between the teller and the parent.

4. Parents of patients with surgically correctable conditions were generally satisfied with the way in which the anomaly was explained to them. Whereas parents did not find it helpful to be shown other patients who had undergone similar surgery, the surgeons' sketches when explaining the nature of the intervention to them were well remembered and appreciated. Due to the hope offered by surgery, parents' initial responses were not overtly emotional. However, a sizeable proportion, notably so in the Hirschsprung's disease group, interpreted the operations as a simple and completely corrective procedure and therefore expected a complete cure to be effected.

A number of measures can be recommended to facilitate understanding of the diagnosis.

(i) It is recommended that other patients not be used as examples to elucidate the outcome of similar surgery.

(ii) In view of its proven success it is suggested that explanations about surgery be further augmented by the use of simple sketches which can be kept by parents for later reference.

(iii) Immediately following the first surgical intervention, certain aspects on the particular anomaly as well as the expectations of surgery should be again explored with the parents by the surgeon. Existing views should then be reconciled to the realities of a specific anomaly. The foregoing can fruitfully be done in liaison with the social worker. In this respect her role should be aimed at identifiying and working with any remaining misconceptions held by the parents.

5. Although the nature and extent of information given to parents at the time of diagnosis could not be verified, the present study identified the parents' feelings of a lack of adequate information. This problem was most frequently raised in the cystic fibrosis group. In the surgically correctable anomaly groups, this was less often experienced. In the latter groups the lack of information pertained more to the delay in conveying the diagnosis in the interim period when tests and investigations were still being done. Furthermore, where one parent had been informed alone, it was found in all the anomaly groups that they had been unable to adequately convey the nature of the diagnosis to the other spouse.

In order to reduce parental anxiety and increase the quality of treatment at this time, it is recommended that

(i) information be given by repetition;

(ii) the volume and the degree of sophistication of the information be geared to the parents' needs;

(iii) the diagnosis be given with the minimum of delay;

(iv) where delay is inevitable, the nature and purpose of tests preceding diagnosis be explained to parents, even in the absence of clearcut results;

(v) the accepted norm in the literature be strictly adhered to, i.e. that both parents be told the diagnosis together.

II. THE CONFRONTATIONAL PHASE: COPING WITH THE ACUTE STAGE OF THE CONGENITAL ANOMALY

At the time of hospitalization new demands were being made on the parents. Many of the problems emanating from the prediagnostic phase, had however still not yet been solved. In addition other major life events, unrelated to the hospital experience, were making demands on the crisis meeting resources of the parents.

A. THE HOSPITAL EXPERIENCE

1. Hospital doctors and parents

(a) Parents were still very uncertain about their credibility with the medical profession. This formed the basis of many problems related to the hospital experience. For instance, in some cases this led parents to deal with seriously sick

infants at home through fear of being told that expert care was not really required. This attitude towards parents was found to be pronounced among the more junior doctors.

(b) The most common complaint by parents was the lack of information about treatment, test results and procedures.

(c) It was found that parents had not been furnished with even the most elementary information, for instance free treatment and the need for physiotherapy in cystic fibrosis.

In view of the above it is recommended that

(i) parents be gradually included in the planning and where appropriate the practical aspects of caring for the child in hospital, which should be accompanied by a progressive build-up of knowledge about the anomaly and its treatment;

(ii) at all times parents be kept abreast of the patients' progress, inter alia by means of explanations on all envisaged tests and treatment as well as feed-back on results;

(iii) parents be furnished with a core of information about the particular anomaly - information which should be repeatedly diseminated by team members who are responsible for the care of the patient, i.e. sisters, physiotherapists and stomatherapists;

(iv) an open door policy on the wards be maintained to facilitate the management of symptoms at home;

(v) an open doctor/patient relationship be encouraged in order to restore parental confidence and facilitate communication which the study has found to be most lacking.

2. Parents and nursing staff

Nursing staff were most frequently in contact with parents and therefore largely determined the nature of the hospital experience for them. Errors in nursing care further reduced parent's confidence in the hospital. This tended to result in more frequent and lengthy visiting by parents in order to monitor patient care. In this way mothers were forced to spend long periods away from the rest of the family.

In view of the above it is recommended that

(i) nursing staff be made aware of their critical role in determining the hospital experience for parents and where possible such incidents as quoted in the study (Chapter 5), be avoided;

(ii) nursing staff be made aware of the emotional trauma which parents were undergoing;

(iii) where nurses do become aware of problems, i.e spending too much time at the hospital, referall to a social worker be made;

(iv) social workers be alert to possible dangers to the

families where mothers spend an inordinate amount of time at hospital.

3. Parents and other hospital staff

The study clearly indicated the positive role of the stomatherapist in restoring parental confidence. This was a direct result of the sensitive and capable way in which she equipped parents to deal with the practical difficulties of the anomaly.

In view of the above it is recommended that

(i) the treatment team recognise the positive role of the stomatherapist in primarily dealing with the practical difficulties of the anomaly, together with the secondary emotional benefits derived by this support to the parents;

(ii) if necessary, the stomatherapist be instrumental in providing any further information to parents where gaps in their information exist.

B. SEEKING A CAUSE AND REASON FOR THE ANOMALY

It was commonly found that parents sought causes for the anomalies. Although on an intellectual level the correct information had been assimilated, wide speculation was present on an emotional level. This was for the most part transient and limited to the acute phase. However, in a small proportion of cases the effects had been pathological. For example, selfblame by mothers, fathers rejecting both mother and child or instances of interfamilial blame was found to be present.

It is therefore recommended that

(i) the chance nature of the surgically correctable anomalies when present be repeatedly explained and reinforced by the various team members involved in management of the patient;

(ii) social workers be alert to the possibility of family conflict or even breakdown as a result of incorrect parental speculation as to possible causes for the anomaly;

(iii) in the acute phase these areas be consciously explored during social work intervention.

C. OTHER MAJOR LIFE EVENTS

The study has shown that a large proportion of families in this sample were dealing with other major life events in addition to the problems posed by the congenital anomaly in the acute stage. This often gave rise to parental behaviour which was interpreted as being "neurotic" or irrational by hospital staff.

It is therefore recommended that

(i) social workers be alert to the fact that many parents will simultaneously be coping with demands which have no direct bearing on the patient and his anomaly;

(ii) these aspects be explored with the parents;

(iii) where excessive life events are found to be present, a mediating role be taken by the social worker to interpret parental behaviour to hospital staff where necessary.

D. BRINGING THE PATIENT HOME

1. Fear of discharge

(a) Some parents (21%) anticipated the day of discharge with fear. This was least likely to be found in the cystic fibrosis group and most often expressed in the oesophageal aresia group where fear of the baby choking was acutely present. On the other hand, parents of patients with Hirschsprung's disease and anorectal malformations soon after discharge began to doubt the wisdom of having the patient at home. These fears were the direct result of difficulties in handling residual symptoms and was more likely to be present patients with periods of lengthy hospitalization.

(b) In all groups fear of discharge was heightened by a feeling of unpreparedness to handle the situation. The study highlighted a number of positive factors which reduced fears of discharge. These included confidence acquired during hospitalization, the open ward policy where emergency admission could be arranged by parents themselves, and the support of a knowledgeable general practitioner in the community.

2. Mother/child estrangement

On the emotional level certain factors militated against an uneventful discharge of patients. Where diagnoses had been made in the neonatal period, there was a greater likelihood of mothers experiencing feelings of estrangement towards the infant. In a lesser proportion evidence was found of the reverse phenomenon: infants feeling estranged from mothers.

3. Death wishes

An additional emotional factor which thwarted the smooth transfer of the infant back into the home was the presence of a death wish towards the patient. Mothers found it difficult to express the fact that these feelings had been present. Although largely limited to the acute phase of the anomaly, in a small proportion of cases these feelings were still acknowledged in the long term adaptational phase.

4. Wishes to have aborted

An important factor slowing down the integration of patients into family life, was the wish of mothers to have aborted in favour of a healthy pregnancy at a later stage.

The findings discussed above clearly indicate the necessity for anticipating post-discharge difficulties. This confirms the expressed need for providing parents with clear-cut instructions on how to deal with the post-discharge problems.

It is therefore recommended that

(i) parents be educated in as many facets as possible of the anomaly during hospitalization of the patients;

(ii) where possible the above be reinforced by written material on dealing with post-discharge difficulties - information which would be equally helpful to caretakers assisting the mother in caring for the patient at home;

(iii) the open ward policy be maintained to deal with emergencies and at the same time allay parents fears;

(iv) the active and practical co-operation of the general practitioner be obtained, especially where parents are distant from major treatment centres;

(v) social work intervention in the acute phase of the anomaly include active examination of emotional difficulties such as mother/child estrangement, death wishes for the patient and the wish to have aborted;

(vi) during visiting hours to the ward mother/child contact be maximised.

III. THE LONG TERM ADAPTATIONAL PHASE: LEARNING TO LIVE WITH THE ANOMALY IN THE COMMUNITY

A. HANDLING SYMPTOMS AT HOME

The study has confirmed the finding in the literature that treatment results, which were regarded as good by hospital staff, were accepted with reluctance by parents. These parental views were influenced by a number of other variables. Because the respective anomaly groups revealed clearly defined differences in those factors influencing adjustment, individual discussion and recommendations are indicated.

1. Cystic fibrosis

It has not been possible to make clearcut recommendations for social work intervention because, firstly, the disease did not follow a chronological sequence of improvement or deterioration, and secondly, the responses of individual families were so widely different. Depending on the current state of a patient's symptoms, parent's moods vaccilated between extreme hopelessness on the one hand to a state of commitment to overcome problems on the other.

A prerequisite for social work intervention is an in-depth knowledge of the various stages and difficulties of the disease as well as a knowledge of the individual families' coping responses. This implies early involvement with families by the social worker in order to deal with the fluctuating and demanding course of the illness through to it's terminal phase. Critical areas warranting social work intervention were identified in the study.

(a) It was found that the fatal nature of the illness was very difficult for the parents to accept or even discuss.

(b) Although the histories revealed that there was heavy investment in patient's home treatment, parents were reluctant to admit to its time-consuming nature.

(c) In some instances a conscious effort was made by parents to keep themselves so busy with treatment that they would not have time to think. This behaviour was interpreted in terms of the phenomenon described in the literature as 'healthy denial'.

(d) Parents in the study overestimated the effects of their home treatment in both positive and negative terms, i.e. when the patient was healthy it was seen as a result of persevering with treatment. Alternatively when the patient was ill this was seen as failure on their part. Deterioration was seldom ascribed to the normal course of the illness.

(e) Parents reacted to the fatal nature of cystic fibrosis by crowding the patient's life with activities.

(f) The increasing age of patients resulted in management becoming progressively more difficult. Issues identified in the study were:

- difficulty in hiding feelings of sadness from the patient
- emotional resistance to treatment in patients
- questions about prognosis and death from patients
- exposure to cruel and insensitive remarks about the illness and death, at school.

In view of the above it is further recommended that

(i) the social worker anticipate and be aware of when parents show a need for assistance in one of the abovementioned areas;

(ii) the social worker serve as an avenue for parents to discuss the fatal nature of cystic fibrosis at a time when they are ready to do so;

(iii) active initiation of such discussion by the social worker preferably be done at the time when it becomes clear that parents are not coping with the demands of the illness;

(iv) the equilibrium gained during phases of healthy denial should not be unnecessarily disturbed in order to explore parents' feelings about death related issues;

(v) the social worker give recognition to the parents for the time consuming nature of home treatment, together with providing an opportunity for ventilation of feelings of anger or futility in this respect;

(vi) where applicable the social worker reinforce to the parents that persistent symptoms are the result of the natural course of the disease and are not the result of failure of their home treatment;

(vii) unless indications to the contrary are found, the crowding of a patient's life be regarded as a normal parental attempt at

compensating for the fatal nature of the illness;

(viii) the social worker be available to share in the parents' feelings of sadness related to the illness;

(ix) the social worker's stress to parents that emotional resistance to treatment is closely linked to normal adolescent challenge of parental authority as well as being a statement of anger and desperation towards having cystic fibrosis;

(x) in addition support be offered to alleviate the concurrent parental anxiety and fear of the patient's death which accompanies any resistance to treatment on the part of the patient;

(xi) the many new issues faced by the adolescent with the cystic fibrosis be handled by means of a combined effort of the management team (clinic) with firstly, the parents and secondly, with the patients individually;

(xii) the above be managed through the medium of groupwork, both with the adolescent patients themselves, as well as with their parents;

(xiii) the social worker assist parents in practical ways in dealing with adverse reactions towards cystic fibrosis at school. (More detailed recommendations pertaining to school-related issues are made in Section V).

All these recommendations will be enhanced by the establishment of open channels of communication both within the families and between the families and the management team.

2. Oesophageal atresia

In contrast to cystic fibrosis hope of increasing improvement in oesophageal atresia patients was a realistic expectation. Some significant features were noted in the study.

(a) By the age of 4 years more than half of these patients no longer suffer from any residual symptoms.

(b) Residual symptoms were limited to choking and the development of food bolus at meal times. Due to the very young age of these patients, parents experienced the symptoms as very acute and frightening.

(c) The nature of residual symptoms were not so embarrasing as to cause social ostracism.

It is therefore recommended that

(i) parents be reassured as to the limited duration of residual symptoms;

(ii) anxiety be further alleviated by practical measures such as

- dietary instruction
- management of bouts of choking and
- maintaining the open door policy;

(ii) social workers be alert to any emotional problems which have not resolved themselves following the disappearance of residual symptoms for example a death wish for the patient extending beyond the acute phase.

3. Hirschsprung's disease and anorectal malformations

In view of the fact that residual symptoms in these 2 anomalies were

very similar, their findings and recommendations will be discussed simultaneously. In both groups the majority of parents mentioned the unpleasant and offensive nature of residual symptoms as an obstacle to easy assimilation of the patient at home. Furthermore the presence of these symptoms extended for a considerably longer period than those of oesophageal atresia. Whereas the latter followed a chronological order in their resolution, symptoms of Hirschsprung's disease and anorectal malformations were influenced by the presence of other factors such as severity of anomaly, social class of parents, parental attitudes and patient motivation towards overcoming the symptoms. Therefore in these two anomalies eventual outcome cannot be ascribed to surgical results alone.

A further complicating factor was the fact that although symptoms had resolved for a small proportion of patients at the time of study, their ages nevertheless varied so widely that recommendations will have to be individualized. The final conclusion was that the anorectal malformation group proportionately exhibited the most problems with residual symptoms followed by the Hirschsprung's disease group. Oesophageal atresia patients manifested the least problems of all the surgically correctable anomalies.

Specific issues identified in the study were the following:

(a) Parents of patients with Hirschsprung's disease tended to expect that surgical intervention would effect a complete cure for the patients. In other words these patients had not anticipated having to deal with the extent of residual symptoms which was found in the sample. The expectations of parents in the anorectal malformation

group on the other hand, were more realistic. It was therefore concluded that although parents had been satisfied at the time, the information given initially did not prepare them for coping with the anomaly in the long term phase.

(b) The majority of parents expressed repugnance for having to perform anal dilatations and therefore discontinued this aspect of treatment at home.

(c) Patient as well as parent difficulties with colostomies together with the socially embarrassing aspects, were adequately illustrated in the text. Where available, the assistance from a stomatherapist was noted to be most helpful.

(d) The unrelenting demands posed by day-to-day management of the patient's toilet habits was a common problem. This was particularly so after the evening meal when toilet training became the centre of family arguments affecting parents, patients and siblings.

(e) In older patients it became more difficult for parents to discuss residual symptoms with them. Both parties were embarrassed and had a feeling that personal privacy was being intruded upon. This was especially so between mothers and male patients.

(f) With the older patients mothers responded either by distancing themselves, i.e. not wanting to know how patients were coping or on the other hand by doing too much for patients.

In view of the above it is recommended that

(i) the extent of information given to parents about the anomaly be reconsidered, especially where Hirschsprung's disease is concerned;

(ii) information be expanded to include more detailed explanation on the nature and duration of residual symptoms as well as the serious nature of the anomaly and it's corrective surgery;

(iii) anal dilatations be done by trained staff in order to relieve parents of these aspects of treatment;

(iv) the availability of stomatherapy services be expanded beyond hospital boundaries to include assistance with colostomies and the performing of anal dilatations in the patient's home;

(v) the helping professions, viz. psychology and social work also take some responsibility for working towards the resolution of residual symptoms in patients, for example by means of techniques from the fields of behavioural therapy and conditioning to enhance patient motivation in this respect;

(vi) social work intervention include both practical and emotionally supportive measures where the handling of residual symptoms effect family life;

(vii) the issues surrounding the adolescent patient with residual symptoms be further researched to evaluate the efficacy of a group approach to dealing with these very personal aspects of symptoms. A stomatherapist would be well placed to run groups of this nature. The above measure would also relieve mothers of some of the responsibility for dealing with this alone.

B. COMMUNITY REACTION AND PARENT ATTITUDE

1. The study has shown that it was important for parents' coping abilities to have a positive and open attitude towards the community. However, the embarrassing nature of residual symptoms caused parents to feel ashamed and sensitive towards community rebuff. This was reflected in the differential proportions of closed attitudes recorded for the different anomaly groups, i.e. oesophageal atresia having the least problems in this respect. Furthermore colostomies were noted to frequently elicit community repulse.

2. In cystic fibrosis closed attitudes were most often associated with parents' views that the disease was not understood by the community which in turn resulted in them being typified as overprotective parents.

In view of the above it is recommended that

(i) every opportunity at educating the community on relevant aspects of congenital anomalies, viz. colostomies be utilized;

(ii) in this respect parents be provided with printed information which they can then diseminate in their immediate environment as the need arises;

(iii) where the social worker becomes aware of an inordinate degree of withdrawal from the community, intervention with these families include active efforts at establishing open attitudes.

C. CONTACT WITH PARENTS OF OTHER PATIENTS

The value of mutual parental support acclaimed in the literature was not borne out in the present study.

Whereas parents of patients with cystic fibrosis had often experienced contact with the parents of other patients, this was seldom the case in the surgically correctable anomaly groups. For the sample as a whole the majority of contacts had been of very temporary nature and not greatly valued by parents. The wide variety of reservations raised by parents indicated that great care should be exercised when introducing parents who had experienced similar difficulties to one another. It was concluded that a potentially valuable source of support had been forfeited because such care had not been exercised.

In view of the above it is recommended that

(i) initial contact be established between parents on an individual basis by the social worker, only in instances where both families are known to her and no contra-indications as listed in the text are found;

(ii) further research directed at how to most fruitfully exploit this avenue of support be undertaken.

IV THE CHILD WITH THE CONGENITAL ANOMALY AND SOME FAMILY RELATIONSHIPS

A. THE MARITAL RELATIONSHIP

The incidence of divorce in the families of the sample was lower than that of the general population, but according to mothers this breakdown was often in some way attributable to the anomaly.

Existing marital difficulties were aggravated by the demands posed by caring for the child with the congenital anomaly. Reported marital strain was noted in half of the families in the cystic fibrosis and anorectal malformation groups and to a lesser extent in the Hirschsprung's disease (43%) and oesophageal atresia groups (17%).

(a) Whereas the physical and emotional absence of fathers were noted as negative factors, fathers' active involvement with management of the anomaly, coupled with emotional support, was most often associated with good or even improved marital relationships. In practical terms those fathers who were able to forego traditional male roles in order to assist with the patient, seemed to have most successfully maintained marital relationships. Furthermore a lack of communication between the spouses about the anomaly, most frequently noted in the cystic fibrosis group, was identified as a negative factor.

(b) No direct link between financial burden and marital disruption was established in the present study. Case studies revealed however that 58% of families in the sample admitted to financial strain with its additional demands on individual family members.

(c) From the study it is clear that areas of strain on the marital relationship cannot be ignored in the overall management of the family of the child with the congenital anomaly and should form an integral part of the supportive services rendered to families.

It is furthermore recommended that

(i) greater involvement of fathers with patient care be facilitated at all times during hospital contact with parents;

(ii) open communication about the anomaly be fostered between spouses during social work intervention;

(iii) social workers remain alert for potential elements of marital strain inter alia lack of spouse support, absence of the father and financial burden as a result of the anomaly.

B. SIBLING RELATIONSHIPS

(a) Negative reactions in siblings were most frequently and intensively found to be present in the cystic fibrosis group. This was followed in declining proportions by the anorectal malformation, Hirschsprung's disease and oesophageal atresia groups. Because of the difficulty in discriminating between normal sibling rivalry and behaviour originating from the psychosocial impact of the anomaly, results are interpreted with caution.

(b) The most common problem was attention-seeking behaviour in siblings. This behaviour was limited to the acute phase of the anomaly and therefore transient in the majority of cases. Simulating cystic fibrosis and regarding treatment or diet as favouritism was found in a small proportion of cases.

(c) The overall concern of mothers about sibling behaviour together with the proportion of "good and helpful" siblings who were carrying responsibilities which were above the age-appropriate level, are areas which warrant intervention.

In view of the above it is recommended that

(i) a more rigorous research design be followed in order to further clearly isolate problem areas of sibling behaviour originating from the presence of a child with a congenital anomaly;

(ii) parents be reassured of the temporary nature of most attention-seeking behaviour;

(iii) social work intervention be geared to relieving mothers of unnecessary concerns as well as dealing with siblings directly in those cases where difficulties exist for them, viz. inter alia being given responsibilities beyond age-appropriate level;

(iv) where possible older siblings be included when factual information about the anomaly is supplied to parents.

V. LIVING WITH A CONGENITAL ANOMALY: THE SCHOOL EXPERIENCE

School attendance posed the first challenge for patients to handle residual symptoms on their own, outside of the protective home environment. The study has shown that the inflexible school system hindered rather than helped the assimilation of patients into the wider community. Very little was done to assist patients' attempts at normal adjustment. Although superficially, there had been no difference between the normal school population and patients, a number of anomaly specific difficulties which detracted from their reaching full potential at school, were identified.

1. Problems specific to the cystic fibrosis group were the following:
   (a) Vital medication such as pancreatic enzymes were either forgotten or not administered correctly by teachers for those primary school patients who had been too young to control their own medication.
   (b) Mothers' attempts to prevent patients from being pushed to their limits with extra-mural activities were interpreted as over-protectiveness by school authorities and were therefore jeopardized.
   (c) Intermittent periods of illness and absence from school resulted in loss of contact with the peer group.
   (d) The eventual death of the patient was poorly handled in the school situation.

2. Post-operative symptoms in patients treated for oesophageal atresia resolved before formal schooling commenced and no difficulties related to this anomaly were found.

3. The difficulties experienced by patients with Hirschsprung's disease and anorectal malformations were found to be of a similar nature and included the following:

   (a) The majority of patients had been subjected to unpleasant incidents at school due to the socially unacceptable nature of symptoms such as staining, smelliness, flatulence, borborygmi and diarrhoea.
   (b) Visiting the toilet as a group at special times, was commonly found in the schools attended by the patients. As these

facilities totally lacked any form of privacy, patients were doubly embarrassed due to their explosive, noisy and smelly stool habits. The communal toilet times were therefore not utilized by patients and gave rise to accidents in the classroom.

(c) Patients were especially vulnerable due to lack of privacy experienced in the locker room and on the sportsfield. For this reason many had attempted but given up participating in extra-mural activities.

(d) For those patients with colostomies the aforementioned problems were especially demanding.

(e) Peer ridicule and ostracism due to the nature of symptoms were often experienced because patients had not been allowed free access to the toilet.

4. Excluding the oesophageal atresia group, some aspects were found to be common to all the anomaly groups.

(a) Underlying most of the abovementioned difficulties at school, was the ignorance of teaching staff about the anomaly.

(b) The study revealed that repeated attempts of parents to explain the anomaly, supply a brochure on the disease or even demonstrate a colostomy, yielded no permanent results. Information tended not to reach significant teaching staff and was therefore of little practical value to the patient.

(c) The almost desperate attempts of patients to conform to the peer group and to make a healthy adjustment was a consistent finding.

(d) The heavy investment of parents to facilitate normal school adjustment for the patient, was well illustrated in the text.

In view of the above it is recommended that

(i) teacher education form the main thrust for dealing with the problems discussed earlier - education which should be commenced at university and training college level;

(ii) knowledge be reinforced by a manual on chronic illnesses which should serve as a standard text of reference in all schools;

(iii) information provided in this way include basic data on

- the anomaly and its residual symptoms
- management and medication
- impact on the pupil
- teacher's role in facilitating school adjustment;

(iv) the above manual be compiled by a Children's Hospital which is knowledgeable about the anomalies and their effects;

(v) teachers in turn be supported by social work services where residual symptoms give rise to adjustment problems;

(vi) practical solutions such as free access to the toilet and positioning the patient close to the door be implemented in order to accommodate the patient in as normal and indiscernable manner as possible;

(vii) where indicated the social worker intervene in order to ensure that the fatal nature of cystic fibrosis be handled with greater sensitivity at school;

(viii) the social worker offer direct support to the patient on how to better cope with the symptoms at school.

The above recommendations would further be enhanced by close cooperation between health, education and social work.

VI. THE LOSS OF A CHILD WITH A CONGENITAL ANOMALY

Findings in this regard must be treated with caution as only a small number of families were involved in this section of the study.

1. The decision to treat the patient

(a) The fathers in this study were well guided by professional staff in reaching a decision whether or not to treat the patient. Mothers seemed to have been involved to a lesser extent in order to ameliorate the trauma at the time immediately following birth. However, the problems arising out of this action were clearly illustrated by the case histories.

(b) Following this decision most parents manifested a need for further emotional support although they had been secure on an intellectual level.

(c) The period of greatest vulnerability was while awaiting the patient's death. At this time decisions about viewing or holding the infant had to be made.

(d) Misconceptions surrounding the death of the patient coupled with religious doubts culminated in guilt feelings in some parents.

In view of the above it is recommended that

(i) all information pertaining to the baby be shared equally with both parents;

(ii) following the decision not to treat, ongoing emotional support be offered by the social worker;

(iii) until proven otherwise by further research, viewing and holding of the baby by parents be advised in order to facilitate mourning following the death of the patient;

(iv) at the same time social workers remain alert as to possible misconceptions surrounding the death of the patient;

(v) religious doubts be explored and if serious, referral for pastoral counselling be made;

(vi) where an anomaly is so severe as to clearly render intervention ineffective, parents not be involved in the decision-making process. In this way a small proportion of parents could be spared the intense emotional trauma experienced in the study.

2. Coping with the death of the patient

(a) The importance of parents being present when patients died was borne out in the study.

(b) In the majority of parents religion played a definite role in influencing their understanding of the death of their child.

(c) Examples of replacing the dead child by means of subsequent pregnancies were given in the text.

(d) In stark contrast to their earlier reticence to discuss the approaching death of the patient, the same parents evidenced an overwhelming need to talk following the death of a cystic fibrosis patient.

(e) In contrast to the foregoing, parents actively avoided talking about the patient's death to friends and

acquaintances if a decision against active intervention had been made in the surgically correctable anomaly groups.

(f) An expressed need for continued contact with the hospital as an avenue of emotional support, was identified in the majority of parents whose children had died. This was especially relevant where post mortems had been done.

In view of the above it is recommended that

(i) social workers actively assist to ensure the presence of parents when patients die, even where this might entail acting as an intermediary between doctors and parents to guarantee adequate indication of imminent death;

(ii) where indicated, the help of ministers of religion be enlisted in order to further facilitate the parents' understanding and acceptance of the patient's death;

(iii) parents be guided in order to prevent overhasty decisions on subsequent pregnancies to replace the dead patient - a role which could be performed by either a social worker or a genetic counsellor;

(iv) following the death of patients the following areas be explored by the social worker

- the need of parents of cystic fibrosis patients to talk about death
- the unresolved feelings surrounding the decision to allow the patient to die
- the unresolved questions of parents following post mortem;

(v) in all instances of death the social worker initiate contact with parents in order to render support during the new crisis faced by the family, i.e. adapting to the loss of the patient.

In conclusion the study has highlighted a few broad issues requiring further investigation.

Firstly, the special problems faced by the adolescent patient with a congenital anomaly have not been acknowledged. Amongst others these include the transition from the Children's Hospital where problems were handled since birth to a hospital for adults where very little is known about the patient and his anomaly.

Secondly, a major shift of emphasis in hospital social work is indicated. This entails movement from dealing with material assistance to becoming involved with the wide spectrum of emotional hardships of parents. This requires early involvement of the social worker to deal with the problems preceding the diagnosis and hospitalization of the patient, while at the same time extending beyond the acute stage of the anomaly to include the long term adaptational phase.

Thirdly, the one factor permeating all the above recommendations and relevant for all staff, is the attitude which they project towards parents. The presence of empathy yielded positive results even in those cases where care was less than optimal. It is therefore clearly indicated that emphasis on attitude should form an integral part of staff training at all levels.

ANNEXURE A

GLOSSARY

ABORAL	away from the mouth
AGANGLIONIC	the absence of ganglion cells
AMNIOCENTESIS	surgical transabdominal perforation of the uterus to obtain amniotic fluid
ANASTOMOSIS	(end-to-end) surgically joining two ends after creating an opening
ASPIRATE	inhale
ATRESIA	congenital closure of a normal tubular organ
BIOPSY	the removal and examination of tissue from the living body, to establish a precise diagnosis
BISTOURY	a long narrow surgical knife used for removing abscesses
BORBORYGMI	the rumbling noises caused by the propulsion of flatus through the intestines
COLOSTOMY	the surgically created opening between the colon and the surface of the body
CONGENITAL	referring to conditions that are present at birth,regardless of their causation
DILATATION	the action of stretching or expanding of a cavity
DISTAL	further from any point, remote; opposed to proximal
DISTENDED	enlarged
ENTEROCOLITIS	inflammation of the small intestine and colon
EXCORIATION	superficial loss of skin,such as by scratching
EXOCRINE GLANDS	glands secreting outwardly via a duct
FISTULA	an abnormal passage (pipe) usually between two internal organs
GASTROSTOMY	surgical creation of an artificial opening into the stomach
HYDRAMNIOS	excess of amniotic fluid
IMPACTION	(feacal) a collection of putty-like or hardened faeces in the rectum or sigmoid
IMPERFORATE	(anus) abnormally closed anus
LIGATION	to tie or bind

ANNEXURE A (cont.)
GLOSSARY

MECONIUM	a dark green material in the intestine of the full-term fetus
MEGACOLON	abnormally large or dilated colon
MYOTOMY	the cutting of a muscle or muscular tissue
NASOGASTRIC	from nose to stomach (usually via the oesophagus)
PERINEUM	the space between the anus and scrotum
PROPHYLACTIC	to prevent or ward off disease
PROXIMAL	nearest, closer to any point
REFLUX	backward or return flow
RESECTION	excision of a portion of an organ
RETENTION	the persistent keeping within the body of matters normally excreted
STRICTURE	decrease in the caliber of a canal
STASIS	an abnormal delay in the passage of intestinal contents
STENOSIS	narrowing of a duct or canal
URETHRA	the canal conveying urine from the bladder to the exterior of the body
VESTIBULAR	pertaining to the cavity at the entrance to a canal
VISCID	sticky or glutinous
VULVA	the region of the external genital organs of the female

ANNEXURE B

TABLE 1

AGE (DAYS) AT THE TIME OF DIAGNOSIS : CYSTIC FIBROSIS PATIENTS

Meconium ileus	2nd affected child	1st affected child
1 2 2 4	7 120 730	75 90 150 169 182 330 730 1030 1277 1460
9	857	5493

X = 2,2 X = 285,6 X = 549,3

ANNEXURE B

TABLE 2

AGE AT THE TIME OF DIAGNOSIS: HIRSCHSPRUNG'S DISEASE PATIENTS (DAYS)

Long segment :	Short segment
1	1
3	2
3	3
4	3
12	3
24	4
30	4
60	11
	20
	21
	49
	60
	60
	135
	150
	150
	210
	270
	425
	730
	1358
137	3669

X = 17,1 X = 174,7

All cases X = 131,2

ANNEXURE B

TABLE 3

AGE AT THE TIME OF DIAGNOSIS: ANORECTAL MALFORMATION PATIENTS (DAYS)

High anomalies :	Low anomalies
All cases diagnosed on first day of life	1 1 1 3 7 90 547 1825
	2475

X = 2 days X=309,38 days (S=640,57)

ANNEXURE B

TABLE 4

DURATION OF FIRST ADMISSION: CYSTIC FIBROSIS PATIENTS (DAYS)

Meconium ileus	2nd affected child	1st affected child
10 29 42 113	5 6 6	0 2 5 5 5 9 10 11 * Unknown
194	17	47

1 = Unknown * Diagnosed by post mortem

X=48,5 days(S=44,96) X=5,67 (S=0,58) X=5,88 (S=3,87)

All cases: X = 17,20 days (S = 28,68)

ANNEXURE B

TABLE 5

DURATION OF FIRST ADMISSION: OESOPHAGEAL ATRESIA (DAYS)

* Type A	Type B	Type C
16	16	42
19	19	90
31	23	Died
60	25	Died
Unknown	30	
	30	
	30	
	32	
	42	
	42	
	61	
	364	
	640	
	685	
126	2039	132

X=31,5(S=20,07) X=145,64(S=236,5) X=66(S=33,94)

All cases: X=114,85 days in hosp on 1st admission (S=202)

* Denotes Waterson's classification

ANNEXURE B

TABLE 6

DURATION OF FIRST ADMISSION: HIRSCHSPRUNG'S DISEASE PATIENTS (DAYS)

Long segment :	Short segment
(1 pt died)	(3 pts unknown)
15	4
22	8
58	10
120	11
125	22
180	22
186	24
	24
	27
	28
	36
	41
	52
	54
	66
	87
	93
	219
	270
706	1098

X=100,86(S=70,58) X=57,79(S=70,91)

All cases: X=69,38 days in hospital (S=72,08)

ANNEXURE B

TABLE 7

DURATION OF FIRST ADMISSION: ANORECTAL MALFORMATION PATIENTS (DAYS)

High anomalies :	Low anomalies
1 pt died 1 unknown 13 26 30 43 60 120 180 191 240 247	 1 2 4 6 11 18 30 32
1150	104

X = 115 days (S=92,4) X = 13 days (S=12,39)

All cases: X=69,67 (S=85,46)

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